

NOTICE TO THE READER

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Dear Parent,

You have received the most devastating and heartbreaking news any parent could ever hear. Your child has Batten Disease.

This is not supposed to happen. When we give birth to a child, we have a dream for our child. It may be that we hope he/she will become a doctor, CEO of a major company, an astronaut, scientist, professional athlete, movie star, or, even president of the United States. Whatever that dream may be, it has been shattered and lost.

You were not given any choice in this matter. Now what? You have been faced with a decision. Either deny this and run away and hide or accept and face the reality. The fact that you have contacted Batten Disease Support and Research Association (BDSRA) is an indication that you have taken a big step in facing the truth that your child has Batten Disease.

You have many questions that need to be answered. “Batten Disease; An Easy to Understand Guide” accompanies this book and is a general information piece for all who seek information about Batten Disease. This book is meant specifically for parents having a child with Batten Disease and attempts to address many of the most difficult questions that you may want answered. Some of the questions and answers are such that you may not want to consider them at this time. Use this book as you see fit.

There are two important things to remember. All is not completely lost. Scientists are making wonderful progress in understanding Batten Disease and are working toward the development of treatments. The road that you will travel is difficult but you need not travel it alone. BDSRA and all the other parents of children with Batten Disease wish to make the trip with you.

A Child with Batten Disease in My Family.

Written by Joan Curran, Psychologist - (edited)

In facing the challenge of being the best parent you can to your child with Batten Disease, as well as to your other children, it can be useful to take stock of your situation. There are three large elements to this, no matter what your circumstances.

1. My child is a child.
2. My child is a child with a disability.
3. My child is a child with Batten Disease.

All of these statements carry complex implications.

My child is a child, first and foremost.

In spite of the overwhelming nature of something like Batten Disease, it is important to try to preserve a sense of your child's own uniqueness and individuality. Your child has a particular temperament and personality, individual likes and dislikes, and lives within a unique set of individual relationships to family and friends. S/he may be outgoing or shy, quite or noisy, active or not. S/he will have particular tastes for music, games, food, clothes, pets, toys, activities. You may see a lot of yourself in your child, including characteristics you like and things of which you are not so proud. You will see traces of other members of your own family. Your child will also have characteristics in common with his/her other parent, again including things you like and things you do not like. Your child will also have traits, habits and attributes for which any antecedent is hard to find. Like every other child, this child is a unique person.

As your child gets older, s/he will be going through the normal childhood sequence of development in many respects. You can expect your child to go through the normal emotional development of any other child, to experience highs and lows, to be naughty and endearing, to have fights and good times with brothers and sisters, with friends and playmates. There may be mood swings, challenges to your authority, increased self-consciousness, and an increased desire for independence. These things occur because your child is a developing human being going through the same process that you went through yourself.

Your child will be emerging as an individual through behaviors which include discovering and testing the limits you set not once, but many times. Like every other child, your child needs to know that these limits are there, and that there is a stable authority figure that can be relied upon to react with consistency, fairness and maturity. S/he needs your attention and your time, not only in relation to physical and medical

needs, but in relation to all the big and little things that are important at the moment. You and your child need to have regular 'special' times together. Childhood years are precious, and in these circumstances, doubly so. Sometimes, we invest our children with our own hopes and inclinations. We say "I've always wanted a son who would come with me to football games" or "I've always wanted a little girl so I could dress her up in pretty clothes". Before we get too carried away, we need to take account of our child's own needs and personality.

You and your child live within a particular set of family relationships, in a particular neighborhood and with your own unique circumstances. In addition to Batten Disease, you must deal with all the other ups and downs of life. If you and your family are going through events which are painful and stressful, such as bereavement, illness, separation, divorce or unemployment, you may find that these things have an impact on your child's sense of security and emotional well-being. Aspects of behavior may be affected. Sleeping or eating may be poor, the child may become clingy or there may be other manifestations of anxiety, and there may be tantrums. These are the normal reactions of children to stress, and they are probably best seen and dealt with as a temporary regression to the behavior and reactions of a younger child. With a child who has Batten Disease, these reactions may be marked or more prolonged.

It is helpful to take time to talk to the child, to answer any questions s/he may have. Children do not always have the words to articulate their feelings very well, but they can feel intensely, nevertheless, and be quite confused. It can be helpful to explain to children what has happened, why things have happened as they did, and what things will be like from now on. When something bad happens, it is not unusual for children to think that it was somehow their fault. If things are not spoken about, this can compound the child's feelings of confusion.

Sometimes even pleasurable events, such as family weddings, travel, or moving to a new house can have elements of uncertainty and fuss, which create stress both for us and our children. For a child who is blind, change and uncertainty can be more disorientating than for a child who can see. If changes and moves are necessary, it is a good idea to prepare the child, for example, by showing him/her a new school or a new house at a quiet time when there is no fuss. At the same time, it is probably best not to begin this preparation process until plans are definite and concrete. You may be thinking of moving to New York City, but to a child, that is as good as saying you are setting off tomorrow.

When changes occur, it is also important to preserve some element of continuity with the past through the presence of a familiar person or through familiar routines and activities. Favorite objects have their function also, and can help to make a new place less strange.

With simple precautions and a little foresight, it should be possible to ensure that your child can participate in many important family events. As well as the sense of

inclusion and belonging which this creates for your child, these landmark events in your family's life can become quite important points of reference in later years, providing a rich source of memories and conversational themes. It is worthwhile retaining some small souvenirs of trips, visits, weddings and so on, as these may be valuable in later years.

My child has a disability.

As the child experiences Batten Disease, it is more like an acquired rather than a congenital disability, in the sense that there are things which s/he used to be able to do and now cannot manage.

For younger children it may be the inability to walk, crawl, speak and even articulate at all. It may seem that the losses have happened overnight. Even then, it is important to continue to assist your child with standing, walking and being active. Encourage him/her to use whatever abilities s/he has at any given time.

For older children this will be a gradual process, but sometimes there is a sudden awareness of failure at some particular task. For any child, this is upsetting, bewildering and frustrating. As a parent, it is natural to want to relieve your child's distress. When your child says "I just can't do this", it is tempting to say "Don't worry, I'll do it for you". Then the job gets done quickly, and everyone moves on to the next thing. However, your child may, in the process, be confirmed in the belief that here is one more thing that someone else must do for him/her. In fact, for quite a lot of activities, whether it is activities of daily living, such as buttering bread or tying shoelaces, sporting activities, or academic work, it is often possible to find an alternative method, an assistive device or a technique which makes it attainable, or else a substitute for whatever is no longer possible. A child who can no longer ride a bicycle may yet get pleasure and exercise from horseback riding. A child who finds it hard to manage at the table can be taught techniques to make the tasks easier, and may find the use of adaptive cutlery and tableware helpful. A child who can no longer read print may learn to use Braille, or to listen to books on tape. This does not mean that learning new methods or adaptations is necessarily easier, and one should not minimize the difficulties faced by the child. However, the restoration of once valued skills, even if only partial or temporary, will do a lot to enhance your child's confidence and feeling of well being, as well as facilitating inclusion in the wider community for as long as possible.

For children with disabilities, life frequently presents more hazards as well as more frustrations. If your child can no longer see, or is subject to seizures, these activities become a lot more problematic. We want our children to be safe, not to get hurt, and, if anything happens, we want to be there. These fears are legitimate, and again, should not be minimized. However, there is also your child's quality of life to consider, and the importance for the whole family of allowing life to proceed as normally as possible, for as long as possible. You will be given advice about safe mobility and about the control and management of seizures. This will not guarantee that mishaps will not occur, but if you prepare well and take all sensible precautions, you should be able to allow your child to enjoy many experiences.

It can be helpful to share your knowledge with a few trusted relatives and friends

in the beginning. Often, people would like to help but do not quite know how to do so. They may be reluctant to intrude or are afraid and so do not offer to look after your child at your home. Sometimes they are nervous about offering to have your child over for parties, play, or short stays because of fears about what to do if an emergency arises. Sometimes you yourself can appear so competent and so in control that the need to offer to help does not occur to them. When the opportunity arises, it can be good to tell those close to you about your child's condition, about what is involved in looking after him/her, and even to ask their advice about ways of retaining their friendships and participating in social activities.

When a child has a disability, it can affect the dynamics of your relationship. Knowing that your child is going through difficult experiences, is missing out on many activities enjoyed by other children, has a lot of hospital appointments and may be struggling developmentally, it can be hard to say no or refuse anything that will bring pleasure. However, not all your child's wishes can be granted all the time, nor is it fair to your child to create the expectation that they can. Like any other child, this child needs to discover the value of deferred pleasure, of give and take, of sharing with others. Saying no to a child with a disability will not damage his/her wellbeing, as long as it is clear that this is a reasonable refusal and that the reasons for it are explained. Your child may not like it, but it will do no lasting damage. On the other hand, continuous indulgence can make a child demanding and petulant. It can limit the willingness of other children to stay around and it can set up an unequal relationship with brothers and sisters which can indeed have damaging consequences for everyone.

Sometimes it is not you as parents who are doing the indulging, but grandparents or members of the extended family. Buying presents or supplying treats can sometimes be the only way relatives can find to express their concern about a child. As long as your own authority is not undermined, a little indulgence on the part of a granny or favorite aunt will not cause any lasting difficulty. On the other hand, if Granny persists in adopting a "poor little thing" line, persists in doing things for the child, which s/he can do perfectly well, and thinks that every whim must be instantly gratified, then it is time to try reach some kind of understanding with her. In situations like this, it is very important for both parents to have a clear position themselves.

Parents worry a lot about the welfare of their other children when they have a child with a major disability. They are conscious of how preoccupied they are, how much attention the special child needs, and how often family life is interrupted. From talking to brothers and sisters of children with disabilities, however, one can be amazed at how ready they are to understand the parents' situation. They realize that Mom and Dad must make a lot of trips to the hospital, that sometimes other people have to be there to get their meals and put them to bed, and that sometimes their brother or sister has special needs which must be seen to, whether it is special rest times, special foods or a larger bedroom.

What brothers and sisters find hard to deal with is if they feel there is an intrinsic

unfairness, for example, if their brother or sister is allowed to be mean to them, or to take or break their property with no redress. Some will complain about an unfair burden of household chores, but will be quite understanding if it is clear that their brother or sister simply cannot do their share. It is striking, too, to hear how angry the able-bodied brothers and sisters can be at the attitudes and ignorance of other people. They can often be much more outspoken than their parents about people who stare, who make foolish remarks or who treat their brother or sister with condescension. There can be strong feelings of sadness at all the things which their brother or sister misses out on, and all the hard things s/he has to endure. It can be helpful for brothers and sisters to meet occasionally with others who are in a similar situation, as many will find that their own friends do not always understand what they have to deal with. It is also very helpful if there is some sympathetic adult, an aunt, uncle, or family friend to whom these children can talk, as sometimes they are unwilling to disclose all their worries and concerns to their own parents for fear of adding to their worries. A relationship like this can prove to be a very valuable resource to the child when times become difficult.

My child has Batten Disease.

It is important for the child who has Batten Disease to maintain as much as possible a sense of normality and regularity, and of continuing with everyday life. At the same time it must be acknowledged that the experience of Batten Disease in a family is not an everyday one, and that there are quite exceptional demands on the family when it occurs. Adjustments, adaptations, and new strategies for doing things can help us along the way for a certain length of time, but a point then comes when new resources are called for, and a different way of coping is required. One parent has said more than once that it is not wise to try to anticipate in advance all the hurdles that have to be met, because one then becomes overwhelmed with the enormity of it all. While it is sensible to have a general idea of the road ahead and the difficulties which will have to be faced, each child's life story will be unique, and there will be variations in the ways in which the condition progresses.

One of the areas which will be affected, but in varying ways, and at varying rates, will be your child's progress in learning. For some children, there may well be a period of years during which they will operate with deteriorating vision but with little other evidence of damage. For children for whom this is the case, it is well worthwhile learning Braille and other skills such as typing, mobility and orientation, just as other blind children do. Particularly if these skills become semi-automatic, they may remain as a source of leisure and relaxation for quite a long time after more marked learning difficulties become apparent.

As the disease progresses, children often have difficulty with short-term memory, which makes the learning of new information more difficult. However, it can be the case that long term memory is better preserved, and that the child's access to previously learned material, such as spellings, history and geography and so on, remains good for quite some time. It can be helpful to make use of this and children can get a sense of achievement by having quizzes and spelling bees organized for them which help to consolidate and use this material.

For parents it can be hard to accept that there is a slowing down and eventually a halt to new learning. It is natural to expect with children, even children who have a very significant learning disability, that there will be a forward progression in the learning of new skills, not the reverse. The child who has Batten Disease will remember a time when s/he was learning new things every day, and may become confused and upset when this is no longer happening. At this point it is important for the child's teachers to have a good understanding of what is involved in Batten Disease, and to realize that they may have to adjust their own goals and expectations for the child. Once the adults in the child's life make this adjustment, alter the demands they have been making, and develop a different set of goals which are more compatible with the child's altered learning capabilities, it can be a great source of relief to everyone.

Sooner or later, difficulties with communication arise. These can be related to

speech. Younger children may never develop a vocabulary of more than a few words or may only develop small sentence structure and are no longer able to develop more expected communication skills. With older children either the volume or the articulation of the child's speech becomes impaired, and makes it difficult to understand, particularly for those who are not very familiar with it. The content of the child's communication can be a problem as well as the difficulties with word finding, repetitive and rambling conversation, and an inability to stop talking. A speech and language therapist can help with some of these difficulties, teaching the child ways of improving voice projection or articulation, or alternative ways of putting things when a particular word proves elusive. There may also be a place for computer-assisted communication, particularly for young people in a more advanced stage of the disease. It is worth remembering that, even though your child may have severe difficulties with expressive language, there may be a much better level of language understanding. It is always worthwhile to talk to your child about gentle, familiar themes, people, places and events.

Parents often report increased emotionalism amongst children who have Batten Disease. This will vary from child to child, but some children may have an exaggerated response to small events becoming disproportionately excited or upset, and hard to calm down. Some children become hung up or obsessed with particular themes, sometimes an event which is due to happen, or a particular interest or hobby. The child may be unreasonably demanding, wanting gratification here and now. The way you deal with these reactions will depend on your judgment about the stage of the disease the child has reached. If your child can communicate fairly well, and can understand a rational explanation, then it is reasonable to expect fairly normal standards of behavior. If your child has become emotionally volatile, then it will be best to reduce the occasions when high levels of excitement are likely to occur, or to avoid too much build-up to upcoming events, trips, etc. Opportunities for regular relaxation are important also, through, for example, music, swimming, gentle exercise, massage, aromatherapy and, for some children, pets.

The more emotionally volatile the child is, the more important it is to try to preserve a regular, calm predictable routine. This type of environment is also best suited to the control and management of seizures. Confrontation is not likely to be helpful at all. There is a place for medication if difficulties become severe. It is also important to be prepared to call on the assistance of other people when you need it.

Every parent of a child with Batten Disease needs a network of supportive people who have an understanding of the condition. It may take some time for you to get used to the idea of someone else sharing in the care of your child, coming in and out of your house, but Batten Disease is a long haul, and you need to conserve your resources in the best interests of your child, your family and your own mental and physical health.

For most human losses there is a sequence of reactions, for example, an initial

denial or rejection of what is happening, bargaining, then slowly an evaluation and exploration of what can be done, and then some kind of acceptance, even though the sense of grief and loss may still be very powerful. Batten Disease involves a heartbreaking series of losses: loss of sight, loss of mobility, loss of learning ability, of speech, and of other bodily functions. There are other losses as well as the child's world becomes curtailed and the opportunities to go out and take part in family and social events become more restricted. Faced with these losses, it is natural for parents to recoil, to feel they don't want to deal with this, to want to blot out the whole thing.

Yet there are almost always practical decisions which have to be made as a direct result of these losses. Your child may need to change schools, to use a wheelchair, a communication aid or other piece of equipment. There may be a need for modifications to the house, or to employ additional assistance at home. Each of these events is a major threshold to be crossed, and each, in its own way, involves both parent and child in a process of letting go. Painful as this process is, once that threshold has been crossed, there is very often a sense of relief in both parent and child. Often, for a period beforehand, there has been stress involved in trying to maintain some facility that is no longer viable.

In dealing with such serial losses, parents may well find that they need help for themselves on a personal level, quite apart from the practical, day to day help they receive in the care of their child. Unique and rare as Batten Disease is, the experience of loss and bereavement is a part of the human condition, and there are many people who are experienced in supporting those who are in grief. Batten Disease is a series of losses and parents experience the grief cycle time after time.

Finally, one more word about brothers and sisters. Although children express their feelings in different ways from adults, they too feel sorrow and loss, and are sometimes frightened and confused by the things they see happening. For example, it can be more frightening to witness a seizure than to have one. Depending on the age of your other child or children, it is important to try to explain what is happening, and what will happen next. Many children who have a very ill brother or sister will be fearful that this may happen to them.

Some children may feel that, because they have, at some point, felt angry with their brother or sister, it is all somehow their fault. Some children feel guilty about their own good health, about the achievements they have, about the good times they enjoy. These feelings may rarely find overt expression, but it is important to take the opportunity to allow the child to talk if and when the occasion arises. Sometimes a good family friend provides a secure support when Mom and Dad are particularly hard pressed.

It is important too that the child who does not have Batten Disease does not feel s/he has to be perfect in every other way, that s/he has to achieve for two, or that any

naughtiness is particularly heinous on their part. Once again, it is important to have additional support outside your immediate family that you can use, so that your other children can enjoy occasional breaks when the care of the ill child becomes intensive. If you can contact other families who are going through the same experience, it can be particularly beneficial for brothers and sisters to meet with one another. This does not have to be a formal, organized affair, though workshops and special activity days are a source of great support, and sometimes form the basis of lasting friendships.

Facing the Challenge

Franklin Roosevelt is said to have stated, “We have nothing to fear but fear itself”. As parents having recently received the diagnosis of Batten Disease it is expected that there is some fear for you. Fear of the future, i.e. “What’s going to happen to my child?”, etc. At this time all is relatively unknown to you.

The unknown is like darkness that closes in around you. **Knowledge** is like a candle whose flame pushes back the darkness and lights the way.

Knowledge about you child’s illness will be the beginning light that helps to overcome the fear. Learn all you can about Batten Disease.

BDSRA has many resources that will help you to learn about Batten Disease. Talk to your mentor and other families who are already on the same road as you. Each piece of written material that you read and each person that you talk to will help you to learn and understand what you and your child are facing.

Batten Disease can be a long wearisome road, but you need not travel the road alone. There are many who will make the journey with you. No one understands better than another parent. BDSRA’s annual conference can be one of the best learning experiences and opportunity to network with other parents. Chapter meetings also provide the same opportunities, but on a smaller scale.

The next section will attempt to answer some of the questions you may have. You may have the answers already. Some of the questions and answers may not be what you want to know or even address at this time. Nothing says you have to read this today, tomorrow, or next week. It is provided only in an attempt to help you.

This next section contains the questions most frequently asked by parents. Many of the questions have no perfect answer. Because of this, some of the answers provided may seem ambiguous or incomplete. At worst the answer will give you some insight or something to consider. Whether or not you agree with the answers, seeking professional advice is always encouraged.

Some Answers to Difficult Questions

Q. Is my child really going to die?

A. Yes, as of today. Tomorrow? We don't know. We hope not. Research is making great strides in understanding Batten Disease. On March 14, 2001 a clinical trial began with a drug to treat Infantile Batten Disease. Today scientists are working toward enzyme and/or gene therapy for Infantile and Late Infantile Batten Disease. Scientists are also making progress with Juvenile Batten Disease.

Q. How do I know that this diagnosis is correct?

A. The only way to know, whether you have doubts about either the doctor or the tests that were performed, is to get a second opinion. It is your right as a parent to do this. BDSRA recommends that there be a confirming diagnosis. This can be done several ways. One way is to find another neurologist with whom to confer or to contact one of the labs known to BDSRA that do the necessary tests. BDSRA can assist with this.

Q. What can I expect in the future for my child? How will this progress?

A. One thing that we can say for certain about Batten Disease, and the thing that can drive every parent crazy, is that it does not treat any two children the same. We can discuss how the disease will progress in general terms but cannot predict just when changes will occur. We are learning that the particular type of defect of the gene can play an important role relative to the progress of the disease. However, there are many other factors that can play a role in the progression that we are not even aware of, i.e., ethnic background, personality, etc.

Q. In general terms, then, what can I expect?

A. Almost all children will experience the same basic progression. The differences will be when the different stages occur and in what order. The children will suffer declining vision loss leading to blindness. Seizures, once begun, will get worse.

Children with Batten Disease will have many forms of seizures including absence (petit mal), grand mal (tonic clonic), drop attacks (atonic), myoclonic, focal (simple partial), and psychomotor (temporal lobe). Not all children will have all the above mentioned types of seizures but will probably have several. Truthfully, the seizures may become increasingly more difficult to control and you may have to try several different anticonvulsants or combinations in order to maintain seizure control. Be aware that anticonvulsants, like any other drug, may have undesirable side effects.

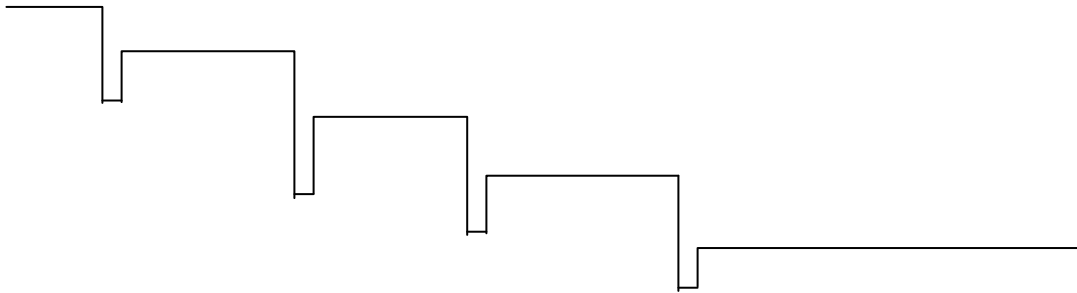
“Clumsiness” will continue to increase until a time when walking becomes difficult and your child will eventually be in a wheelchair. Gross motor skills will continue to deteriorate.

Speech/words will deteriorate until gone. Eating and drinking will be compromised, also.

Cognitive functioning will also deteriorate. Memory loss, possible behavioral

problems, sleeplessness and sleep disturbances may occur. Children with Batten Disease begin a normal development after birth but, depending upon the form of Batten Disease, will “peak” and begin a regression.

With all forms of Batten Disease, one can expect a continuing series of losses, rebounds and plateaus. This is shown graphically below.



We do not always know what may precipitate a drop, how much of a rebound there may be or how long a plateau will last. Drops are sometimes caused by an illness such as the flu and sometimes it is the progression of the disease itself. The plateaus are also unpredictable and can last up to a year or more.

- Q. What did I do that caused my child to have Batten Disease?
A. You did not do anything. Batten Disease is not caused. It is a genetically inherited disorder. Both parents must be carriers of the same defective gene for the possibility of there being an affected child. Please refer to the section in *Batten Disease: An Easy to Understand Guide* that explains genetic inheritance. Unfortunately, neither you nor the other parent of your child was aware that you are carriers. No one is aware until they have an affected child. Genetic specialists today say that everyone carries four or five defective genes. So, do not blame yourself. You did not know.
- Q. Will my older child/ren develop Batten Disease? If the one child has Late Infantile, can my older child/ren develop Juvenile?
A. No! First, if your older child/ren do not show symptoms by this time, they are clear. Second, each form of Batten Disease is the result of separate genes. The gene for Infantile is different from the gene for Late Infantile, which in turn, is different from Juvenile, which is different from the gene for the Adult. If father and mother each carry the gene that causes Late Infantile, then all affected children would have Late Infantile.
- Q. Should I tell my child that s/he has Batten Disease and s/he is going to die?
A. Many people advocate telling the child everything. Many do not. Some parents do and some do not. There is no right answer to this question. Only you can make

that decision. Whatever you feel is the right thing to do is the right answer. Never let someone else make that decision for you. Your decision is your decision to make, no one else.

Some older children, especially those with Juvenile Batten Disease, are more aware that things are not as they should be. They realize that things they were once able to do are no longer doable. They realize that something bad is happening to them. Some of these children ask for an explanation. Often a simple answer will suffice. However, some children become very adamant about wanting to know. You may be backed into a corner and have to provide an answer. What you say and how you say it depends on how much you believe your child will understand or accept. Perhaps you may not have to tell all at one time. Perhaps some explanation over a period of time will be sufficient. Unfortunately, there are no magic words for this situation. If your child believes in Heaven and angels, that may help.

Q. Should I tell the other children in the family?

A. Telling the other children will depend a great deal on their ages. Younger children often accept the fact that their brother/sister is simply “sick”. Older children will sooner or later begin to ask very pointed questions. Eventually they will have to be told. However, again, you do not necessarily need to explain everything at once. You know how much they will understand and be able to handle. When questions do arise, “sugarcoating” is the most obvious thing to do. Children are generally smarter and more tuned in than we may think. They expect honest answers.

Q. Should I have my younger child/ren tested to determine if they have Batten Disease?

A. That is entirely up to you. If you suspect that a younger child may have symptoms of Batten Disease then perhaps you should. You would find out for certain one way or the other. If younger child/ren are not showing any symptoms then there is no apparent hurry. Either way, time will eventually tell. If a younger child has the disease, time will eventually cause the onset of the first symptoms. The question is: Do you want to know now or later? Some parents have their younger child/ren tested soon after the first child is diagnosed. As some of them have explained, “Not knowing is like watching a time bomb and wondering if it is real or when it may go off.” Testing can either confirm or defuse your worst fears.

Q. What do I tell other family members, such as grandparents?

A. Tell them everything, tell them nothing or tell what you want. Today may not be the day you want to explain Batten Disease. Maybe tomorrow or maybe next week. They will have to be told sooner or later, but that is up to you. You can also

refer them to the internet or share the information that you receive from BDSRA and let them learn at their own pace.

Q. Once I/we have told our parents, how do I/we get them to understand that the defective gene came from both sides of the family?

A. Batten Disease is known as an Autosomal Recessive Inherited disease. That means that both father and mother have to carry the defective gene which in turn means that they have had to inherit the gene from either their father or mother. Auto-somal means that the disease does not affect just males or just females. There is a chart (Appendix 1) that shows in detail how recessive inheritance works. Hopefully the chart will help to explain. The chart shows that father's father is a carrier and that mother's mother is a carrier. It could be opposite or it could be both grandfathers or both grandmothers are the carriers. Regardless, it is only an example of recessive inheritance.

Q. My spouse is in denial. Is this normal? What can I do to help?

A. Denial is normal to a point. Whenever there is a traumatic experience, loss or death in a person's life they will experience the "grief cycle" which includes denial, bargaining, anger and eventually acceptance. A diagnosis of Batten Disease is a traumatic experience for a parent and other family members as well. The diagnosis and disorder are difficult to accept, thus the denial. Many children early in the progression of the disorder do not really appear to have anything wrong, thus the denial that there IS anything wrong.

Moving through denial and onto acceptance is something that must take its own time and course. No two people do this the same way or in the same time frame. Understanding and patience by the spouse is important. However, if the person remains in denial for an excessive length of time or the denial is having adverse affects on either themselves or the family, counseling may be in order.

Q. Many family members and friends offer to "help" but I do not know what to say. Also, many say they would like to help but do not know what to do.

A. Never be afraid to ask. The beginning time is the best time to get other family members and friends involved. As time passes the demands on your time become more and more. Let people whom you trust come and stay with your child for a period of time so that you can get out and do your shopping, go to the bank, post office or wherever. Setting a ritual of a particular day or evening that you and your spouse get out together, for even a short time, becomes important. Your child also enjoys having others come in and play, watch videos, etc. Perhaps letting someone take another child to a game or practice would be helpful. Use your imagination and do not be afraid to let others help.

Q. I'm terrified of seizures. What can I do to get over the terror?

A. Seizures, especially grand mals, are terrifying. Parents never really get used to seizures. They just learn what to do when a seizure occurs. You can learn about seizure recognition and first aid from different sources such as your local hospital's neurology department, American Red Cross, and by talking to other

parents. The Epilepsy Foundation of America often puts on training sessions in which you can participate.

Q. I hear a lot today about diet and nutrition. Is there anything special I should do with my child's diet?

A. Diet and nutrition are always important. A normal healthy diet is always recommended. You may have read in some material about Batten Disease that the diet should be low in Vitamin A. This is no longer true. A normal intake of Vitamin A is okay. Just like any other person, your child's diet should not be overly high in Vitamin A is all. As time passes and the disease progresses you may experience problems getting your child to eat what you wish him/her to eat. At this point getting your child to eat period can become a feat. Now is the time to let him/her eat whatever he/she will eat. You can always supplement the diet with nutritious additives. One thing to be aware of is weight. Too much weight is not good for anyone, but as mobility decreases for a child with Batten Disease your assistance in helping to move him/her in and out of the car, for example, will become more difficult if your child is overweight.

Q. What about supplements?

A. Supplementing your child's diet with vitamins may be a good idea. Talk to your child's physician about this. For many years supplementing with Vitamin E, B6, B12 and selenium has been a method of attempting to reduce the build up of storage material in the cells. For some children this seems to help and for others it does not seem to have much effect. Other supplements currently being given are carnitine and fish oil. If you wish more information about these, please call BDSRA.

There is a great movement toward using natural supplements such as valerian root, ginkgo, St. John's Wort, etc. We have nothing to show that any of these have a positive or negative effect with Batten Disease. Many parents are giving their child a natural supplement. However, if you talk to different parents you'll find that each has their child on a different regimen. BDSRA has no position on this subject. Also keep in mind that natural, over-the-counter supplements are not regulated as to quality, etc.

Q. What are some things I should be considering now?

A. Dealing with Batten Disease is a "team" effort. It will eventually take a combined effort of many people to help you. Among those are your child's pediatrician, pediatric neurologist, dentist, teacher, and family members. Keeping these people informed and educated of what you and others are doing and of any changes that may take place with your child is a worthwhile effort that keeps each informed.

If your child is now on any anticonvulsants or other drugs, post a list showing times and dosages in a place where everyone will know its location. This eliminates guesswork. Carry a note card with the same information with you at all times. Your spouse should also carry the same information.

Develop a checklist for drugs. Whenever a dosage has been administered check it off. This prevents either not giving or giving the medication/s twice.

See also the next section entitled "Records".

Records.....Records.....Records

As was previously mentioned in the Q & A, record keeping is essential in many ways. Keep in a separate file every piece of paper or information that you receive.

Never let any original letter, test result or other information get away from you. If

someone needs something from your records, make photocopies to send away. This way you always have the original on hand to make more photocopies if needed. Photocopies of photocopies of photocopies eventually reduces legibility to near zero.

Keep a log book. There are so many things to remember and memory can become confused after a time. Keeping a log book helps you to pinpoint changes, etc. Things to keep in the log book include:

Visits to the doctor, dentist, ophthalmologist, hospital and any type of therapy, etc. Record the date, reason and outcome.

Seizure activity including date, time, type of seizure and duration. Charting all of this from your record book may show a pattern that will lead to changes in times of administering medicines or dosages. It may also reveal a “trigger” that starts a seizure. A sample chart is shown as Appendix 2.

Changes of drugs, i.e. drugs added or discontinued, changes in dosages, reactions, etc. This means any drug including over-the-counter drugs.

Whenever a new drug is administered keep a record of what happens for a few days. This may pinpoint a reaction such as mood changes, excessive sleeping, hyperactivity, physical changes in gait/mobility or speech and behavior. Be aware that any child can have a reaction to any drug. Keeping a record of these things can be just as important as recording when a drug has the desired affect.

Telephone calls that have anything to do with your child. Record time, date, and subject of call along with the outcome. Remember to get the name of the person with whom you are speaking. This is especially important if it is someone with your insurance company or a social service such as SSI or Medicaid.

Changes in behavior can be important to record. Often you will find patterns, especially with girls as they reach puberty. Be aware that with puberty it is not uncommon to see an increase in behavior problems and/or an increase in seizure activity as a girl approaches her menstrual cycle. Strangely enough, some parents have also reported an increase in behavior problems and/or seizure activity during a full moon. Also record what may have been going on around your child prior to any significant change in behavior. Often a “trigger” to some behaviors can be found.

Record results of any test, i.e. blood tests, MRI, CAT Scan, etc. If possible get a copy for your file.

A NOTEBOOK THAT TRAVELS BETWEEN YOU AND YOUR CHILD’S TEACHER/S CAN BE IMPORTANT.

Finish the day with a general, short summary of the day. Good day, bad day, etc.

It may seem that all of this record keeping is a long, time consuming exercise.
Do it! Down the road you'll be glad you did. Guaranteed.

Effects of a Terminal Illness on a Sibling Relationship

by Robbie Hatchett

Many different variables are considered when thinking about how a young tree grows to be a strong oak. Quality of soil, weather of the surrounding region, closeness of the roots of other trees, and human interaction are all factors that affect the growth of a tree. If all factors are perfect, there is a good possibility that the tree will grow to its

fullest potential. However, if even one factor has problems, the tree may go through several trials in order to grow. If the tree conquers these problems, its roots will be firmly established. The description of this tree is very much like the process that a child goes through when growing up. If the child has the "perfect" family, there is a good possibility that he will grow to be a fine man. However, most everyone experiences various trials while growing up. The question that determines how positively a child will grow up is, "How will the child deal with trials?" These trials come in various forms. One test that most children experience is developing a successful relationship with their siblings. This challenge can be difficult for many families. However, for families that have "special" children, this task can take on an increased level of difficulty.

Anyone who has had a brother or sister probably knows the advantages and difficulties that occur in a "typical" sibling relationship. Siblings can take on many roles toward each other. The first that is generally taken is playmate. As siblings grow, teacher, friend, companion, and protector are all roles that siblings could hold for each other. However, there is no universal way to describe a sibling relationship, since all have their own individual characteristics. In many families the siblings spend more time with each other than they do with their parents. Many children learn some of life's first lessons from their siblings. Sharing, competition, rivalry, compromising, and fighting are all actions that many siblings engage in while young. Though this sibling relationship is just between the children, parents can have a huge influence on the relationship. Parents must strive to meet the needs of their child, but can sometimes only meet one child's needs at a time. Problems can begin to occur if parents show one child more attention over another. Feelings of jealousy, envy, and hostility can begin to appear in the child who is shown less attention.

What happens when a sibling relationship is not "typical"? Sometimes an uncontrollable factor is placed on a sibling relationship, and a parent may not be able to carry out the "typical" actions of a parent because of extra stress placed on the family. Parents who have a child with a terminal illness must deal with this type situation. Though a family (family = mother, father, diagnosed child [child with a terminal illness], and healthy child) with a terminally ill child might want to carry on normal family actions, many unavoidable twists and turns can be added that, like a tree, will either restrain a family or make it grow stronger. In most families, there are three relationships: parent/child 1 (diagnosed child), parent/child 2 (healthy child), and child 1/child 2. With the appearance of a terminal illness, these relationships can have totally new characteristics.

A child with a terminal illness can pose many new challenges for parents. Instead of looking forward to seeing their child grow up and excel in different areas, they are faced with knowing that their child will not be able to lead what many would consider a full life. Many concerns and stresses are now added to the parents. Continual care of the ill child, worry about the life span of the child, and guilt of causing the disease are all factors that can be present in everyday life. The parent must, of course, strive to meet the special needs of the diagnosed child. Many times these needs will be above and beyond what the typical family is accustomed to providing. Many of the parents must consider

care of the ill child before any decision is made. Activities must revolve around the diagnosed child now. So, where does this leave the relationship between the healthy child and the parents?

Children have many different feelings instilled in them by their parents. Love, support, attention, and safety are all things that need to be present to provide secure self-esteem for a child. However, when there is a terminally ill child in the family, this can put a strain on interactions between parents and the healthy sibling. First, parents are often pre-occupied with the necessary care the diagnosed child requires. Next, parents might be tired from caring for the diagnosed child and often do not have enough energy to spend on the healthy sibling. As the relationship between the diagnosed child and the parents grow because of their time spent together, the parent/healthy child relationship can weaken. Furthermore, if parents are not able to fulfill all the demands of caring for the diagnosed child, more responsibility may be expected from the healthy sibling. This could pose opportunities for the family to work together and grow closer, or it could push the healthy sibling farther away. The factor that may now have the greatest importance is the need for communication. The diagnosed child will affect everyone in the family in an individual way. The parents cannot know how the healthy child is dealing with the situation and may have no idea of the turmoil that is going on inside the healthy child if there is not good communication between them.

The presence of a terminal illness obviously can affect the relationship between parents and their children. However, the relationship between siblings can be affected, also. Many new challenges are now present in the relationship. Instead of playing ball together, fighting, or seeing who can run the fastest, new ways of bonding must be found. Sometimes, taking care of or doing chores for the diagnosed child might become a new way of bonding for the healthy child. A terminal illness has many effects on the outward actions of all family members.

Growing up in today's society can be very trying. Add the fact that a person has a sibling with a terminal illness, and the amount of strain and stress during childhood can greatly increase. Having a diagnosed brother or sister can affect the social, psychological, and emotional development of a child. The sibling will have to adjust to the fact that the diagnosed child will require extra time, attention, money, and support throughout his whole life. This causes many healthy siblings to have a tough time making the necessary adjustments, and many undesired feelings can occur.

The terminally ill child will need more attention than the healthy child. This fact can cause many negative reactions in the healthy child. First, attention seeking behaviors may begin to arise in search of finding ways to gain the needed attention. Disruptive behavior may begin to occur more frequently. The healthy child would hope that these actions would force the parents to turn their attention away from the diagnosed child and towards the healthy child. Likewise, tantrums become a possible action because this too would steal attention from the diagnosed child. Next, the continual thought that the diagnosed child is getting more attention may cause the healthy child to develop feelings of jealousy towards the diagnosed child. This jealousy can lead to even stronger feelings

such as hostility. When a thief steals something that a person thinks is rightfully his, the victim will probably become mad. Such is the case when a healthy child feels the diagnosed child is stealing attention. Since the siblings in this family probably cannot fight as other brothers and sisters do, the healthy sibling must store his anger and release it through these different behaviors.

The thing that children learn from the attention of their parents is the reassurance that they are being supported in a stable relationship of love. With the lack of this reassurance, many reactions are possible. First, the healthy child might become "clingy" to the parents. This is done by always showing the need for physical contact with the parents. This physical contact, though instigated by the sibling, would produce the feeling of being loved. Next, a sibling might try to develop his own handicap. This handicap, known as a psychosomatic pain, would be developed by the imagination of the healthy child because he realizes that the diagnosed child receives attention on account of his/her illness. Also, the possibility of promiscuity may become a very real problem. Love is one of the greatest needs that a person has, and if a person cannot get that love from the family they will search in other places. These actions might receive some attention from the parents, but it is not the true, loving attention that every child needs.

As the healthy child grows, he may have many new feelings that pertain to the child with the illness. Many of these feelings come subconsciously. First, feeling guilty because the healthy sibling is spared the disease that plagued the diagnosed child is common. Second, anger is usually present at some time in a healthy sibling's life. The sibling can release this anger towards God, fate, the parents, the diagnosed child, or even at himself. Third, healthy siblings might feel that they were the cause of the disease. Fourth, a sense of fear is sometimes present because the healthy sibling might think that he could somehow contract the disease. Next, the healthy sibling might feel an added pressure to excel in various activities. Since the diagnosed child might not be able to participate in many "normal" activities, the father might have a stronger desire for the healthy child to exceed in various sports, leadership roles, etc. Sixth, an increased feeling of "parentification", taking on various roles of a parent, is present in many healthy siblings who have a diagnosed sibling who has to be cared for on a constant basis. Seventh, healthy siblings may be embarrassed to be seen with the diagnosed child in public because of the negative ways society often views people with disabilities. This, in turn, could have various effects on the social life of many healthy siblings. The fear that people might think they are odd because of the condition of their diagnosed sibling might cause a healthy sibling to shy away from close relationships. Next, the fear of how a boyfriend/girlfriend might think about the diagnosed child is ever present. Ninth, healthy siblings might feel left out when dealing with the diagnosed child. A need for information about the disease, its cures, and the condition of the diagnosed child might always be on a sibling's mind. However, parents sometimes feel that it might be best to keep this damaging information away from the healthy children. Finally, many siblings have a fear because of the uncertainty of the future of the diagnosed child. Knowing that a child has a terminal illness can put added pressure on healthy siblings because they know that their brother/sister's days are numbered. This can affect decisions regarding college, careers, and marriage that a sibling has to make.

There are many factors to be considered when thinking about how a healthy child will react to the terminal illness of a sibling, including differences in age, gender of the children, number of other siblings, and the nature of the disability. Next, if a healthy sibling has already moved out of the house when the child is diagnosed, the siblings might not have as close a relationship as siblings who live in the same house. Also, the disease might be so bad that the diagnosed child is not able to live at home which would take away many of the daily stresses that other healthy siblings experience. However, the biggest factor that can determine how a sibling might react to difficulties caused by the diagnosed child is the way that the parents handle the situation between the siblings. To have a successful family in this situation, parents must strive to know the needs of the healthy sibling. Since the healthy sibling can face many different feelings inside, the parents need to know how to satisfy those feelings. If the parents know that the healthy sibling is jealous because of the attention that the diagnosed child receives, then they can make sure that something is done to ensure that the healthy sibling knows the love that the parents have for him. Knowing the needs of the healthy child could help to avoid many of the negative actions and feelings that are exhibited because of the diagnosed child.

Despite the many stresses and difficulties that can be added to a healthy sibling because of the illness of a brother or sister, many lessons can be learned that would shape a healthy sibling in a way that people in a "typical" sibling relationship might not understand. First, a deeper and quicker maturity is often characteristic of a healthy sibling. Second, through dealing with a child whose days are numbered and through experiencing many family struggles, a healthy sibling can develop a sense of what the truly important things are in life. Third, a deeper appreciation of family and health can be apparent in the lives of healthy siblings. Fourth, healthy siblings are usually more sensitive towards humanity and charitable with their actions. Increased tolerance of differences and compassion are other valuable lessons that can be learned from living with a child who is "different" in the world's eyes. Through successfully dealing with the strenuous trials that a terminally ill child can place on a family, a healthy sibling can grow a rich loyalty towards the diagnosed child and the rest of the family. Finally, the most beneficial feeling that can be gained from living with a child with such a terminal illness is the pride that a healthy sibling can have in a diagnosed child. This pride might take a while to develop, but after it has peaked, it is a joy that cannot be taken away. The smallest things that a diagnosed child achieves can now bring joy to everyone's life.

Many negatives and positives can be present in a sibling relationship that involves a terminal illness. The positives can outweigh the negatives for a healthy sibling. I am a "healthy sibling". At age 4, my brother began showing the first signs that made my family realize he had a terminal illness. He began to lose his sight and was totally blind at age 8. Seizures, retardation, loss of the ability to talk understandably, and the loss of ability to walk were next. Stephen, my brother, was diagnosed with Batten Disease. It is a neuro-logical disorder that is fatal to every child who has the disease. I went through most every stage psychologists report that a healthy sibling goes through. I had times where I did not get the attention I needed. I had times where I had to do various

caregiving jobs for my brother that are not present in most typical sibling relationships. I had times that I was mad at God, my parents, or anything else that I could blame for the disease. I had times of guilt because I was spared of the illness. I had and still have fears about where to go to school and what kind of career to pursue because of the uncertainty of the lifespan of my brother. However, these times have made me see the world through a clearer set of eyes. Through living with a blind child, he has made me see the truly important things of life. All the positive characteristics that were mentioned are all qualities I feel I possess. However, words cannot explain the joy that I find through the experiences with my brother and through the life that my brother leads. I am proud of every accomplishment, no matter how small they may seem, that he achieves.

“When life throws you lemons, make lemonade.” This statement could be applied to many different types of struggles people face. It seems especially applicable to people who have a sibling with a terminal illness. Many roadblocks can occur in a healthy child's life because of the uncontrollable situation. As young healthy siblings grow into adults, they have two directions they can choose. Either the healthy siblings can allow the struggles and disadvantages to consume their lives, or they can look at the situation as an opportunity to live with and learn from a very special child. The parents can also help lead healthy siblings in the right direction by striving to understand the difficulties they face. If the parents can make the necessary adjustments, the healthy child has an even greater opportunity to see the bright side of this situation.

10 Tips for Family Caregivers

1. Choose to take charge of your life, and don't let your loved one's illness or disability always take center stage.
2. Remember to be good to yourself. Love, honor and value yourself.
You're doing a very hard job and you deserve quality time, just for you.
3. Watch out for signs of depression, and don't delay in getting

professional help when you need it.

4. When people offer to help, accept the offer and suggest specific things they can do.
5. Educate yourself about your loved one's condition. Information is empowering.
6. There is a difference between caring and doing. Be open to technologies and ideas that promote your loved one's independence.
7. Trust your instincts. Most of the time they'll lead you in the right direction.
8. Grieve for your losses, and then allow yourself to dream new dreams.
9. Stand up for your rights as a caregiver and a citizen.
10. Seek support from other caregivers. There is great strength in knowing you are not alone.