

ACKNOWLEDGEMENT

We would like to extend our sincere thanks to

SPECIAL PEOPLE IN NEED,

*the organization whose generous grant in 1998
made the original publication of this book possible.*

This is the fourth edition.

~ Revised 2011 ~

Notice to the Reader and User of this Guide:

All material in this Guide is provided for information purposes only. Although the Batten Disease Support and Research Association (BDSRA) makes every reasonable effort to assure the accuracy of the information contained in this Guide, BDSRA is not engaged in rendering medical or other professional services and advice. BDSRA does not guarantee or warrant that the information in the Guide is complete, correct, current, or applicable to every situation. BDSRA disclaims all warranties, express or implied, concerning this Guide and the information contained herein. If medical or other expert assistance is required, the services of a competent professional should be obtained.

Batten Disease Support and Research Association
All rights reserved.

Table of Contents

What is Batten Disease?	6
History of Neuronal Ceroid Lipofuscinosis	7
What are the forms of NCL/Batten disease?	7
How many people have these disorders?	8
What causes these diseases?	8
How are these disorders diagnosed?	8
What research is being done?	9
Is there any treatment?	10
What is happening to the student?	10
Vision Information for the Regular Classroom Teacher	13
Muscular Control	15
Fine motor	15
Gross motor	15
Physical Therapy and Batten Disease	17
Normal motor development	17
Implication of Batten disease	17
Physical and motor symptomology	17
Physical therapy interventions	18
Maintaining gross motor skills while maximizing function	18
Provide support and security	19
Prevent injuries and deformities	19
Social Interaction	20
Cognition	21
Speech/Language	24
Communication	26
Finding an alternative	26
Calendar systems or schedules	30
Daily calendars	30
Calendar box hints	33
Early experience with objects	34
Experience books	35
Summary of Best Practice	36
Individualized Education Plan (IEP) Goals	38

Educational Strategies for Children with Infantile and Late Infantile Batten Disease	40
Establish a ‘yes/no’	40
Turn-taking	40
The ‘Little Room’	41
Resonance board	43
Calendar boxes	43
Augmentative devices	45
Experience book	45
Behavior Management	46
Behaviors related to the disease	46
Assess the environment	47
Functional behavioral assessment and analysis	49
Individualized Education Program (IEP)	52
Present levels of academic achievement and functional performance	52
Annual goals and short-term objectives or benchmarks	53
IEP participants	54
Realistic expectations	55
Parental Participation	58
Placement in Special Education	59
Inclusion	60
The move to a more restrictive educational placement	60
A need for continuity	61
Related Services	62
Transportation	62
Speech-language pathology	62
Audiology services	63
Psychological services	63
Physical and occupational therapy	63
Recreation	64
Social work and counseling services	64
Orientation and mobility services	64
Vision services	64
Medical services	65
Procedural Safeguards	66
Notes for the School Nurse	67
Health care plans	67
Make Time Count!	69
How to Contact Us	70
References	71

Illustrations

Figure 1: Pressure switch for listening to books-on-tape.	23
Figure 2: Augmentative device for expressive communication.	28
Figure 3: Augmentative with objects attached for tactile recognition.	29
Figure 4: Calendar box system with 'finished' side.	32
Figure 5: Lilli Nielsen's 'Little Room.'	42
Figure 6: Lilli Nielsen's resonance board.	44

Teach and Be Taught

A Guide to Teaching Students with Batten Disease

A challenge awaits you. That challenge is to educate your student who has Batten disease, and this guide will help you improve the quality of life for your student and assist the family in accepting the challenges that they face. Batten disease offers little hope at this time in the medical arena however, the progression of the disease is generally predictable. Utilizing the predictability of the disease, this guide provides the knowledge base to planning a quality educational program for the student with Batten disease. It is vital to know the progression and end results of the disease to make time count concerning education for the student with Batten disease. What you teach now can benefit and empower the student even in the final stages of the disease. Recognize, though, that there will be individual differences among students, requiring education to be individualized according to students' individual needs.

What is Batten Disease?

Batten disease is named after the British pediatrician who first described the disease in 1903. The juvenile form is the most common form of a group of disorders called Neuronal Ceroid Lipofuscinosis (or NCLs). Although Batten disease is usually regarded as the juvenile form of NCL, it has now become the term to describe all forms of NCL in North America. The basic cause, progression, and outcomes are the same. The forms of NCL are classified by age of onset but are all genetically different.

Generally, children are the ones affected with this disease, although there is a rare form of NCL that affects adults. Depending upon the type or form of Batten disease, the age of onset will vary. Generally speaking, children are born well and reach some or many developmental milestones in the first few years of life. Onset is defined as the time when the illness manifests itself, and onset may be very subtle in the beginning. Initial symptoms may be seizures, diminishing vision, or clumsiness. These are common symptoms in many diseases/disorders. Because Batten disease is so rare, the initial diagnosis is often incorrect (i.e., epilepsy, retinitis pigmentosa, macular degeneration, developmental delay, or mental retardation). However, as the disease progresses, it soon becomes obvious that these diagnoses are incorrect and the search for the real diagnosis begins. Batten disease is, at best, extremely difficult to diagnosis.

Over time, affected children suffer mental impairment, worsening seizures, and progressive loss of sight and motor skills. Eventually, children with Batten disease become blind, bedridden, unable to communicate, and unable to eat. Batten disease is always FATAL. This disease does not affect any two children in exactly the same way. The disease knows no timetable, and it cannot be predicted when these things will occur. The severity of these aspects also cannot be predicted. The same is true concerning the age of death. Batten disease is not contagious or, at this time, preventable.

History of Neuronal Ceroid Lipofuscinosis

The first probable instances of this condition were reported in 1826 by Dr. Christian Stengel in a Norwegian medical journal. Dr. Stengel described four affected siblings in a small mining community in Norway. Although no pathological studies were performed on these children, the clinical descriptions are so succinct that the diagnosis of the Spielmeyer-Sjogren (juvenile) type is fully justified.

More fundamental observations were reported by F. E. Batten in 1903 and by Vogt in 1905, who performed extensive clinicopathological studies on several families. Retrospectively, these papers disclose that the authors grouped together different types of the syndrome.

In 1913-14, M. Bielschowsky delineated the late infantile form of NCL. However, all forms were still thought to belong in the group of "familial amaurotic idiocies," of which Tay-Sachs was the prototype. Subsequently, it was shown by Santavuori and Haltia that an infantile form of NCL exists, which Zeman and Dyken had included with the Jansky-Bielschowsky type.

What are the forms of NCL/Batten disease?

There are ten types/forms of NCL, of which eight have been identified in the USA and Canada. All but one are childhood disorders starting anywhere between six months and ten years of age. The remaining form is an adult onset and is quite rare. The symptoms are similar, but they become apparent at different ages and progress at different rates.

Infantile NCL (Santavuori-Haltia disease) typically begins between six months and two years of age and progresses rapidly. Affected children fail to thrive and have abnormally small heads (microcephaly). Also typical are short, sharp muscle contractions called myoclonic jerks. Initial signs of this disorder include delayed psychomotor development with progressive deterioration, other motor disorders, and/or seizures. The infantile form has the most rapid progression, and children live only into their mid-childhood years.

Late infantile NCL (Jansky-Bielschowsky disease) typically begins between ages two and four. The common early signs are loss of muscle coordination (ataxia), seizures, and progressive mental deterioration. This form also progresses rapidly and ends in death between about ages eight and twelve.

Variant late infantile NCL (CLN6) typically begins about age three and has the same onset symptoms of the "classic" late infantile. As with late infantile, the disease progresses rapidly, and ends in death between about age ten to twelve.

Juvenile NCL (Batten disease or Batten-Spielmeyer-Vogt disease) typically begins between the ages of five and eight years of age. The common early signs are progressive vision loss, seizures, and ataxia or clumsiness. This form progresses less rapidly and ends in death in the late teens to early twenties, although some may live into their thirties.

Adult NCL (Kuf's disease or Parry's disease) generally begins before the age of forty, causes milder symptoms that progress slowly, and does not cause blindness. Although age of death is variable among affected individuals, this form shortens life expectancy.

NOTE: There are, however, rare occurrences when an individual will not fit the "norms" and may or may not experience the loss of motor control, sight, and mental abilities as fast as others affected with the same form of Batten disease.

How many people have these disorders?

Batten disease and other forms of NCL are considered to be relatively rare, occurring in an estimated 2 to 4 of every 100,000 births in the United States. The disease has been identified worldwide. Although NCLs are classified as rare diseases, they often strike more than one person in families that carry the defective gene.

What causes these diseases?

Symptoms of Batten disease and other NCLs are linked to a buildup of substances called lipopigments in the body's tissues. These lipopigments are made up of fats and proteins. Their name comes from the technical word "lipo," which is short for "lipid" or "fat," and from the term pigment, used because lipopigments take on a greenish-yellow color when viewed under an ultraviolet light microscope. The lipopigments build up in cells of the brain, eyes, skin, muscle, and many other tissues. Inside the cells, these pigments form deposits with distinctive shapes that can be seen under an electron microscope. Some look like half-moons (or commas) and are called curvilinear bodies. Others look like fingerprints and are called fingerprint inclusion bodies. Still others resemble gravel (or sand) and are called granular osmiophilic deposits (GRODS). These deposits are what doctors look for when they examine a skin sample to diagnose Batten disease.

The exact cause of Batten disease is not known. It is known that the disease causes death of neurons (i.e., brain and nerve cells). However, what causes neuron death is not known. The buildup of lipopigments that happens in the case of Batten disease is not the cause of cell death. What is known is that in Infantile and Late Infantile Batten Disease the buildup of lipopigments is the result of missing enzymes whose job is to rid the cell of these lipopigments. The cause of the buildup for juvenile and other forms is not known.

How are these disorders diagnosed?

Because vision loss is often an early sign, Batten disease may be first suspected during an eye exam. An eye doctor can detect a loss of cells within the eye that occurs in the three childhood forms of NCL. However, because such cell loss occurs in other eye diseases, Batten disease cannot be diagnosed by this sign alone. Often, an eye specialist or other physician who suspects NCL refers the child to a neurologist, a doctor who specializes in diseases of the brain and nervous system. The neurologist needs the patient's medical history and information from various laboratory tests in order to diagnose NCL. Diagnostic tests used for NCLs include:

Blood or urine tests: Tests that detect abnormalities that may indicate Batten disease. For example, elevated levels of a chemical called dolichol are found in the urine of many NCL patients.

Skin or tissue sampling: The doctor examines a small piece of tissue under an electron microscope. The powerful magnification of the microscope helps the doctor detect typical NCL deposits, common in skin cells and sweat glands.

Electroencephalogram/EEG: An EEG records electrical currents inside the brain through special patches placed on the scalp. Doctors are able to detect patterns in the brain's electrical activity that suggests a patient has seizures.

Electrical studies of the eyes: Visual-evoked responses (VER) and electro-retinograms (ERG), detect various eye problems common in childhood NCLs.

Brain scans: Allows doctors to look for changes in the brain's appearance. The most commonly used imaging technique is computed tomography (CT), which uses x-rays and a computer to create a sophisticated picture of the brain's tissues and structures. CT scans may reveal brain areas that are decaying in NCL patients. A second imaging technique is magnetic resonance imaging (MRI), which uses a combination of magnetic fields and radio waves to create a picture of the brain.

Enzyme assay: Infantile and late infantile are missing a known lysosomal enzyme. Enzyme assay determines if the enzymes are missing.

DNA testing: Nine of the ten genes are known. The gene for adult onset NCL or Kuf's disease has not yet been identified. DNA testing can identify whether an individual has one of the defective genes and is the best method of determining which form of Batten disease affects the individual.

What research is being done?

Within the Federal Government, the focal point for research on Batten disease and other neurogenetic disorders is the National Institute of Neurological Disorders and Stroke (NINDS). The NINDS, a part of the National Institutes of Health (NIH), is responsible for supporting and conducting research on the brain and central nervous system. The Batten Disease Support and Research Association also promotes, assists, and funds research.

Through the work of dedicated scientific teams, the search for the genetic cause of NCLs is gathering speed.

In September 1995, the International Batten Disease Consortium announced the identification of the gene for the juvenile form of Batten disease. The specific gene, CLN3, located on Chromosome 16, has a deletion or piece missing. This gene defect accounts for 73% of all cases of Juvenile Batten Disease. The remainder is the result of other defects of the same gene.

Also, in 1995, scientists in Finland announced the identification of the gene responsible for the infantile form of Batten disease. This gene, CLN1, is located on Chromosome 1.

In September 1997, scientists at the Robert Woods Johnson Medical School and the Institute for Basic Research, NY, announced the identification of the gene for the “classic” late infantile form of Batten disease. The gene, CLN2, is located on chromosome 11.

The genes for six of the remaining seven forms of Batten disease have also been identified. The gene associated with the adult onset remains undiscovered. Identification of the specific genes for infantile, late infantile, variant late infantile, juvenile, and other forms of Batten disease has led to the development of DNA diagnostics, carrier and prenatal tests, and enzyme assays, also for diagnostic and prenatal testing.

Is there any treatment?

As yet, no specific treatment is known that can halt or reverse the symptoms of Batten disease or other NCLs. Scientists are working on several different approaches to the development of therapies. It is not yet known if any of these therapies will stop the disease.

Seizures are sometimes reduced or controlled with anticonvulsant drugs, and other medical problems are treated appropriately as they arise.

Support and encouragement help children and families cope with the profound disability and losses caused by NCLs, while scientists pursue medical research that could someday yield an effective treatment.

What is happening to the student?

Batten disease causes death of neurons (brain cells). As the neurons die, more and more symptoms of the disease become apparent. Seizures begin and continue to intensify as time passes. The child may suffer from different types of seizures, such as absence (petit mal), tonic clonic (grand mal), atonic (drop), myoclonic (sudden jerks) and complex partial (psychomotor). Seizure activity varies from child to child. A key to understanding Batten disease is to understand that the illness knows no specific pattern or time schedule. One child may have one type of seizure, and another child may have two different types of seizures. One child may have an onset of seizures at age five, another child at age nine, and still another child at age thirteen. The teacher and staff must have an understanding of what seizures are and what to do if one happens. Again, remember you may see a seizure that does not follow the typical types of seizures generally known. One thing is for certain; **DO NOT PLACE ANYTHING IN THE CHILD’S MOUTH DURING A SEIZURE!** Just make sure the child is as safe and comfortable as possible during the duration of the seizure.

As previously noted, one of the initial symptoms of Batten disease is the beginning loss of vision. The vision loss is not reversible and will lead to total blindness. Color blindness and loss of central vision occurs first. The child eventually looks at objects, people, etc. out of the corner of the eye. This peripheral vision usually lasts for awhile. When the peripheral vision is lost the child is left with light/dark perception. Before total blindness occurs there is a period when the totality of darkness will come and go.

An additional symptom of Batten disease is clumsiness. The disease affects the parts of the brain that deal with mobility, and clumsiness manifests soon after or during the onset of the illness. This ataxia worsens as the illness progresses and mobility will continue to decline. Batten disease results in an eventual total loss of mobility.

In addition, as the brain cells die and the disease progresses there is loss of the ability to eat, loss of speech, loss of continence, and loss of cognizance.

There are two things to remember. First, there is no timetable when these things will happen. We can say in general that these things will occur but not when or even in what order they will happen. Second, the progression is slow and will happen over many years. Batten disease is compared to the teeth of a circular saw. There are times when the child will suffer a loss or losses, a rebound, and then a plateau or level time. No one can predict when a drop or loss will occur and what may precipitate it. No one can say how much of a rebound there will be. The plateaus may last for weeks or even months before another drop or loss is experienced. The plateaus are welcome and valuable periods that allow the child, family and teacher time to adjust.

Children with Batten disease will attain developmental milestones on or ahead of schedule in their early years. Crawling, sitting, walking, word development, numbers, tying shoes, talking, sentence development, and so on, right into math, language, and writing are normal with children who have the juvenile form of Batten disease.

The learning curve of a child with Batten disease, as compared to a “normal” child, shows they are alike until after the onset of the disease. After onset, the learning curve of the child with Batten disease begins to slow. As time advances, the curve begins to flatten and will eventually “peak.”

When a child with Batten disease begins to peak, some areas of learning will stop while others continue to advance, although slowly. This process fluctuates. Students who are able to perform tasks, later in the same day may not be able to perform the same tasks, and still later are able to accomplish it properly. It is when this peak is reached that a teacher needs to consider a reverse trend in thinking that will correspond to the reversal that the child will undergo. The child should be challenged to continue to learn, but expectations need not be as high as would be for a child that is not experiencing neurologic regression.

Major regression begins once the child has passed beyond the peak. The downhill decline begins and does not end. The child loses things in a reverse order. In other words, the last thing learned will generally be the first loss. As the regression continues, the child will lose short-term memory and retain long-term memory for a long time.

When the regression begins the teacher becomes challenged. Teachers are taught to teach. How do you deal with “unlearning?” The following is an illustration how one teacher solved the problem. A girl with Batten disease loved spelling. She began with the usual easy words (e.g., dog, cat, etc.) and progressed to where she was spelling large difficult words. When she began to peak she would be able to correctly spell her list of words and a short time later not be able to spell any of them. Given an hour or two break, she would once again correctly spell all the words but later not be able to spell them. It was back and forth. Once she really began to regress the teacher adjusted her word list by going backwards and giving her easier words. She eventually ended up back at the simple words of dog and cat. The teacher heaped praise on her, and the student glowed in the praise and felt a sense of accomplishment.

Vision Information for the Regular Education Classroom Teacher

With the progressive degenerative condition due to Batten disease, loss of vision will occur. The visual loss, at first, may not be apparent, but the signs of visual loss will increase in the classroom setting. The teacher needs to be aware of the following symptoms:

1. Changes in visual acuity.
2. Inconsistent and erratic performance of far point visual tasks such as seeing the writing on the chalkboard or a poster across the room from the student's desk.
3. Squinting and shifting to focus on near point visual tasks such as reading or while trying to assemble a project directly in front of the student.
4. Asking more frequent questions or asking for verbal repetition of instructions regarding the student's own performance when vision is part of the learning activity.
5. Gradual shift in participation in classroom activities that require vision.
6. Changes in physical proximity to the sources of instruction or activities. The student may initially seek out a closer location and later not know to move closer to those sources for information.

The child may not always see other's toys/obstacles that may be on the floor as the visual field diminishes. A cane or a helping hand may be of great assistance to the child. In addition, interventions that the classroom teacher should consider for the student with Batten disease include:

1. Teach/speak in close proximity to the student.
2. Preferential seating for activities where usable vision will enhance learning such as sitting closer to the speaker or nearer the chalkboard.
3. Providing more light at the student's work areas to promote the use of residual vision. Intensify the light source and mix the types of lighting, trying to include natural light as well as florescent and incandescent lighting.
4. Give clear, large or enlarged print examples and orally discuss those examples, asking the student questions that will reflect the learner's understanding of the presented concepts and ideas.
5. Use clearly contrasting materials in visual presentations. Dark or bold ink on off-white or cream colored paper stands out more than the pastel shades of colored pencils or papers. Ask the student which ink and paper stand out the best.
6. Use original dark ink material rather than penciled work for the student's ease in reading.
7. Directly show graphs, charts, diagrams, pictures, and other visual models to the student with verbal discussion and accompanying comprehension questions.
8. When using typed print, use a good quality print that is legible to the reader.
9. When using computer generated materials, select laser print for clarity.
10. Assign a "buddy" and have the students work in cooperative pairs, repeating the instructions orally to each other and especially to the student with Batten disease.
11. Increase the amount of auditory instruction along with written directions.

12. Have students examine three dimensional learning materials often, such as a relief map of their state, feeling landforms on a world globe, and studying time by manipulating unifix cubes to represent minutes on a large clock face.
13. Ask the student to be part of demonstrations and part of role playing to encourage more active participation as long as possible.
14. Enlarge the print on presented materials, even if this means using two pages for presenting the content of a single page.
15. Use manipulative objects for the student to study. Relate the objects directly to the learning tasks at hand.
16. Discuss new information posted in the room and teach their locations (i.e., a bulletin board, the door, on the wall, or at a table), so the student can learn the new information.
17. Allow the student to choose from a variety of writing instruments.
18. Reinforce often with verbal praise when the student attempts to actively learn in the classroom.
19. Incorporate the student's known information into the beginning discussion of unknown information.
20. Use a print enlarger when visual documents need enlarging.
21. Ask the vision specialist for other specific suggestions that may be appropriate for the student.

Discuss with the parents how well the student handles a variety of visual tasks in the classroom. Indicate the successful as well as the less successful situations. Inquire of the parents if they have found any particularly effective techniques to deal with the vision loss in the home that might also be tried in your classroom. As the student's vision will continue to change, ask the parents to update you on any visits to their vision specialist. Above all, remember this is a team effort of parents and professionals where sharing and communicating needs to take place much more frequently than with other students and their families. Remind them that you are there to be part of the support team to help their child. Your care is valued and appreciated by the families.

Muscular Control

The area of muscular control encompasses both fine and gross motor abilities. Fine motor skills include handwriting, dressing, block building, and many other skills that require finger dexterity and coordination. Gross motor skills include walking, running, playing sports, and many other skills that use the large muscle groups of the arms and legs. Muscular control is a focus of the educational process due to the regression of these skills as the disease progresses.

Fine motor

Fine motor activities become increasingly difficult for the student with Batten disease. The student knows what he or she wants to do but often cannot make the fingers perform. This periodic breakdown between perception of the brain and the actual motor act often causes frustration that results in the student acting out inappropriately in many cases. Therefore, it is the teacher's responsibility to read the student and recognize the level at which to challenge and when to simply nurture. For example, a particular young student loved to build with Lego blocks. He built intricate structures and was very proud of his creations. However, as his abilities declined, he engaged in this activity less and less. A very wise teacher recognized what was happening and adapted the student's block building activity by providing bigger blocks that were easier to put together. The student once again happily engages in his favorite activity and creates elaborate, bigger, structures. This teacher read her student and recognized how and when to challenge him.

Braille, a skill often taught to students with visual impairments, becomes more of a fine motor difficulty than a cognitive task for the student with Batten disease. The numbness that occurs in the fingers of such students impairs their ability to interpret the small raised dots. The philosophy in Holland concerning Braille is to teach it if the student shows an interest in learning this form of communication. Otherwise, if no interest is shown, do not waste time trying to teach Braille. This skill will diminish early, and time can be more wisely spent teaching skills that will endure the disease process. Ultimately, read your student. Focus on the fine motor activities that he or she enjoys and adapt those activities to allow the student to be successful.

Gross motor

Gross motor activities focus on mobility, endurance, and strength. Two very different schools of thought have arisen in the area of gross motor skills. One, provide massive amounts of therapy to maintain gross motor abilities for as long as possible. On the other hand, the school of thought from Holland is that it is sadistic to prolong something that is slipping away. Both philosophies show merit. Therefore, this decision becomes a personal choice of the family. However, when the physical therapy becomes very stressful to the student it is time to back away from that activity. Undue stress may increase seizure activity. This trade-off is not worth pursuing. The student must find enjoyment in any physical activity.

Again, read your student when balancing activities that address gross motor skills. For example, one young girl loved her gymnastics class that she attended once a week. She also received physical therapy twice a week. As therapy became too stressful, the mother decided to decrease physical therapy to once a week and increase gymnastics class to twice a week. A student that is losing his/her ability to walk may find enjoyment by practicing walking in a swimming pool. The buoyancy of the water enables the student to continue to walk when the ability out of the pool is no longer possible. The swimming pool will also stimulate circulation and allow the student a better sense of where his/her body is in space. The key here is to read your student carefully. Remember, physical stress on the child may increase frustration and could lead to increased seizure activity. Therefore, intervention in this area must be enjoyable to the student. Also, allow the student to determine his or her level of participation on a day to day basis, acknowledging that some days the student will feel less confident to fully participate.

Physical Therapy and Batten Disease

Normal motor development

It has been said, “*We learn to move and move to learn.*” Normal motor development, or more specifically the acquisition of gross motor skills, is observed through the development of “motor milestones” (i.e., head-up, roll, sit-up, creep, stand, & walk). These motor milestones, along with the brain's developing ability to organize and process sensory stimuli, enable an individual to respond, move about, orient, and function in various and changing environments.

The body's motor responses to environmental changes are known as 'postural' and 'equilibrium reactions' and are observed in adjustments made in posture, protective reactions, balance, and equilibrium. Physical therapists are able to measure the quality of these movements and make adjustments to provide information regarding the individual's safety, motor dependence or independence, functional/developmental level, motor progression/regression status, and intervention strategies.

The motor system does not develop in isolation. Closely integrated is the sensory system, which organizes sensations that in turn elicit a motor response. This sensory motor system in the early years is the foundation for more complex skill functions such as writing, reading, behavior and social skills. As a child with Batten disease develops, many sensory motor skills will be affected, some skills will be lost, and others need be relearned in an adapted way.

Implications of Batten Disease

Important considerations must be noted in dealing with children with Batten disease. The regression and devastating effects of Batten disease are not always predictable; the intensity and timetable of physical decline and involvement is unpredictable. Unknown individual factors are the order and degree of losses and each child's tolerance level to these losses. As with other diseases or injury to the brain, symptomology and ramifications of Batten disease can be widespread and extensive. Deterioration is documented in motor, sensory, communicative, behavioral, cognitive and psychosocial functioning. This is often observed in vagueness or declines in self-awareness, problem solving, judgment, attention and information processing, including short-term memory and thought organization.

Physical and motor symptomology

Because of the marked deteriorating nature of Batten disease, thorough evaluations, reassessments, accurate and measurable record keeping and documentation are highly important.

The motor disorders observed with Batten disease at first may be subtle and inconsistent. However, as the disease progresses, symptomology increases as does the intensity of the disorders. Deterioration may be evidenced in one or more of the following:

1. Diminished postural mechanism (loss of head righting, protective reactions);
2. Loss of balance/equilibrium (clumsiness, stumbling, falling);
3. Deteriorating posture (bowed-rigid, linear, lacks rotation, increased flexion of extremities, knees bent, 'sinks' into gravity);
4. Changes in gait pattern (wider base, toeing-in, shuffle gait, crouched-gravitational insecurity)
5. Changes in breathing patterns (shallow, short);
6. Abnormal muscle tone
 - hypertonic (usually extremities-spastic/rigid, lacks full range-of-motion)
 - hypotonic (usually of trunk with decreased stability);
7. Ataxia (lack of graded control of movement of trunk and extremities);
8. Extraneous-involuntary movements;
9. Muscle weakness and atrophy;
10. Decreased facial expression (mask-like);
11. Difficulty initiating actions (swallowing, speech, transfers);
12. Tremors (often combined with ataxia and spasticity, limits gross & fine motor);
13. Apraxia (difficulty in motor planning, affects fine/gross/oral motor tasks/swallow);
14. Dysarthria (collection of oral motor disorders-speech, chewing, swallowing); and
15. Decreased motor planning, orientation, localization, language-word retrieval.

Physical therapy interventions

The role of physical therapy in the school system is evaluating, identifying, and developing motor goals that promote and facilitate maintenance of gross motor skills, maximize the quality of movement and function, and provide support through adaptive equipment when necessary. The ultimate goal is to enhance the student's well-being and continued learning, even if through different modes and at different paces.

The following recommendations are not prioritized in order of importance or need. These are only a few suggestions for therapy and many times activities will yield benefits for more than one area. Again, the underlying goal is to enhance the child's well-being and quality of life while working within the child's ability and tolerance levels.

Maintaining gross motor skills while maximizing function

1. Provide opportunities for mobility (i.e., ball, pool, or mat exercises).
2. Provide opportunities for deep and increased breathing.
3. Motivate with enjoyable activities. Combine motor movements with auditory stimuli (i.e., stationary bike while listening to music).
4. Maintain touch as tolerated. Use firm contact and avoid light touch if not tolerated.
5. Maintain proper body alignment, especially in sitting and laying.
6. Continue standing and weight bearing. Use adaptive equipment as needed.
7. Provide a peaceful, safe, positive, and non-threatening environment.

Provide support and security

1. Provide safety bars for physical support transfers and ambulation.
2. Utilize splints and braces as needed to enable weight bearing and mobility.
3. Utilize adaptive equipment to provide secure, well-aligned sitting, standing, and ambulation.
4. Support wheelchair use for ease of travel for child and family as needed.
5. Provide recommendations for 'in-home' physical management.
6. Always provide moral support to student and family.

Prevent injuries and deformities

1. Plan ahead! Allow additional time and avoid hurrying for all activities.
2. Provide a familiar, secure, barrier free environment for safety and ease of exploring.
3. Facilitate mobility and weight shifting as long as possible (i.e., bed mobility, reaching, lay-to-sit, roll, etc.).
4. Adapt activities of daily living as needed (i.e., foods easy to chew and swallow, straw for ease in drinking, tub/shower seat with spray hose, elevated toilet seat, safety bars for all transfers, bed rail, avoid throw rugs).
5. Insure a well-aligned posture in positions.

The importance of active movement and mobility cannot be over emphasized! However, use caution and delete or adapt activities that cause stress.

It has been researched and reported that sensations and the motor responses to sensations act as nourishment to the brain. However, due to each child's individual tolerance level and abilities, the role of the educator is to encourage, facilitate, and assist in active mobility as long as it is possible and enjoyable for the child. When active participation becomes stressful, intervention strategies need to be changed with more emphasis on comfort, support, assistance, and adaptive equipment.

A full team approach with professionals, parents and the student working together will accomplish the most in promoting the greatest potential for quality-of-life for the child with Batten disease.

Social Interaction

Social interaction becomes a vital issue when questions arise concerning the educational placement. Many families have a difficult time removing their child from a regular education setting to a more restrictive educational placement. However, do not allow the inclusion panacea to sacrifice the well-being of the student. Remember, students with Batten disease know what is happening to them. They know what they once could do that their peers are still able to do. This knowledge often surfaces as problematic behaviors. The student may be trying to tell those around him or her that it is extremely painful to be educated with peers that continue to progress while he or she struggles. Perhaps this is the time to place the student in a more restrictive educational placement. Social interaction with familiar peers can continue but on a different level, such as peer tutoring or mainstreaming for subjects that the student still enjoys participating in with his or her non-disabled peers. Once again, it is vital to read the student.

It is important that the child remain socially involved at his or her own comfort level. Even after the loss of sight, the child will remain very aware of the environment and activities around him or her. Physical contact and talking with the child is very important to maintain a connectedness with the environment. Reading to the child and describing what activities are happening also enhances his or her quality of life. In addition, discuss with the parents some of the activities the child enjoyed or experienced prior to the onset of the symptoms. You will then be able to say things such as: "Remember green is the color of the grass," and "Here is Mickey Mouse. You remember him from your trip to Disneyland." **ALWAYS** talk with the child as you would anyone else that age regardless of the lack of vision or communication skills.

The child wants to maintain independence. However, as the illness progresses, the child has to be moved towards dependence. Teaching tactics will eventually change from teaching new things to teaching and maintaining daily living skills.

Cognition

Dementia is defined as the loss of cognizance. Batten disease can bring a whole new meaning to the word. For some children the dementia process can be extreme in that it can produce severe behavioral problems. These may range from severe outbursts of anger, uncontrollable emotions, striking out, hitting, biting, hysterical laughter, heart wrenching sobbing, and hallucinations. At its worst, the psychotic behavior can manifest itself in self-abuse. Like many other aspects of Batten disease, how the dementia process may affect a child is not predictable. Some children have no problems, while others have severe problems. The remainder is somewhere in between. The dementia process is also the least understood and most difficult with which to deal.

In the moderate to severe form the dementia can become almost intolerable. The uncontrolled behavior may be an almost impossible situation, especially if the child becomes vocal. *Echolalia* is repetitive speaking. The child may ask a question over and over and over and although you may give an answer, the child continues to ask it again and again. Usually, this period is relatively short lived and will pass. Often, the most difficult time is when the child's thought process becomes disrupted. The outbursts can come at any time without warning and usually for no apparent reason. No amount of logical resolution will work to deter what the child may have on his or her mind at the time. It is especially important to be aware that these children still understand the difference between appropriate and inappropriate behavior. They often cannot help what is happening. Hallucinations usually seem to take on two aspects: 1) persons who are obviously not present, or 2) terrible things such as snakes, spiders and monsters. We know that all of these are not real but to the children they are very real and threatening.

All of this can be disruptive to the classroom setting. You, the teacher, your aides, and the other classmates need to understand that the child is usually not able to control what is happening. There often is no good resolution to this disruptiveness. Often times it is trial and error as to what will work best to bring quiet and order to the classroom. **Remember**, usual behavior modification techniques are typically not effective.

There are a couple of strategies that may help. First, as the dementia process takes hold the child does better on a predictable schedule. In particular, an object schedule helps the student that has difficulties handling changes to routine or normalcy. When the dementia is severe, anticipation and disappointment may also cause problems. For example, the class is going on a field trip to the zoo in four days. During the intervening time, the anticipation of this event may build and build to a bursting point. If, for some reason, the field trip is canceled the child having Batten disease is likely to have problems, as the reason for the cancellation will not be understood. In addition, the anticipation itself may cause problems just because it is anticipation that cannot be controlled. If you know that anticipation and/or disappointment are becoming a problem, consider waiting to tell the child about an event until it is about to happen. This may appear contradictory to the previous statement relative to sudden changes in schedule. However, changes that lead to pleasurable activities may not cause any difficulty. You may have to weigh one against the other.

Another strategy that is often helpful is playing a child's favorite music or video-tape during an outburst or when there is difficulty. The children typically have favorite music or videos that they listen to over and over. Also, let the child hold a favorite object that represents this activity

throughout the activity. This strategy gives the child something to fidget with as well as help the child focus on what is happening. Be assured that the period of dementia will pass. No one can predict how long it will last, but what is usually noted is a gradual abating of the dementia until it is completely gone. Unfortunately it also signals the end of many other functions.

Several drugs have been tried to ease the dementia problems the children experience. Sometimes they help, sometimes they help for awhile, sometimes they have an opposite effect than expected, and sometimes the drugs have no effect at all. Like everything else with Batten disease there is no single drug or method that will work for every child.

A key point in all of this is that the child is aware of what is happening to him or her. The child is aware of the continual slowing of abilities and eventual losses that take place cognitively, emotionally, and physically. This awareness can exacerbate the dementia. Frustration levels can be exceedingly high for the child when he or she cannot retrieve words, move about as easily, use hands and fingers, and express him or herself as well as before.

The dementia that occurs with Batten disease appears somewhat different from other neurologic regressive diseases. Long-term memory stays quite intact. This fact is very significant to you as the teacher of a student with Batten disease. Focus instruction on areas that will be important and meaningful to the student in the future. For example, teach object cues that correspond with activities that the student enjoys even when the student still has verbal communication skills. Objects taught early are more likely to remain in long-term memory and will allow the student to access them when the verbal skills are gone. This area is covered in depth in the section on communication.

Students with Batten disease love to reminisce about past experiences. Discuss past events with them often to help secure significant events in their long-term memory. These memories bring comfort and stability to a life that is facing chaos. Make 'memory books' or 'experience books' of significant events told in story form along with a tactile picture. The written stories allow caregivers and others to talk about the events and to understand the child's attempts at discussing them. The tactile representations enhance the recall process for the child.

The most important aspect of cognition is for the teachers and caregivers to realize that the student with Batten disease possesses a great deal of intelligence even when he or she cannot access this knowledge. Never underestimate this intelligence. Formal assessment suggests a decrease in IQ. However, this decrease may be due only to a decrease in performance, rather than an actual decrease in IQ. What these students actually know and understand is unknown to us and untestable by standard measures. Regardless, assume that your student knows and understands everything and never talk down to him or her. Always treat your student with the same dignity and respect that a normal peer enjoys. Emily, a fifteen-year-old girl with Batten disease, would become extremely agitated when her teacher did not understand this and would sing nursery rhymes to comfort her. This only enraged Emily further. Once the teacher understood, she was able to make progress with Emily. They were often found in discussions about boys, philosophy, friends, family, and even death. These discussions pleased Emily, calmed her, and stimulated further intellectual thoughts. Emily would smile rather than cry. The agitation was gone.

In Holland, the day for students with Batten disease begins with the teacher reading the current events from the newspaper. The students appear happy and content as they intently listen to what is happening in the world around them. Accessing the written word brings back memories and comforts the student with Batten disease. Trevor is a fourteen-year-old boy with Batten disease. Trevor has little control over his jerky movements due to the seizure activity and neurologic damage resulting from Batten disease. However, he loves listening to books on tape, and he will work extremely hard at controlling his excess movements when activating the tape recorder with a switch (Fig.1). Even in the advanced stage of the disease Trevor knows that releasing the pressure on the switch will result in the reader stopping as well. Favorite books from Trevor's past bring him great joy as he actively and alertly accesses them in this way.

Figure 1: Pressure switch for listening to books-on-tape.

Speech/Language

The following is a list of characteristics found in the early stages of Batten disease. A diagnosis of retinitis pigmentosa in a young child raises a red flag for professionals working with the child. Monitor this child closely. Watch for:

- A plateau of academic skills
- Hesitations, repetitions, and stuttering-like speech
- A somewhat awkward gait, with the knee or foot turned slightly inward
- Blank stares that may only last a few seconds

The Speech/Language suggestions that follow may prove beneficial with a child in the early stages of Batten disease.

1. Early on, one can tell that the child's ability to comprehend language supersedes their ability to express language. Thus, one may find ways to help this child express his or her thoughts; verbally, with sign language, and/or with augmentative communication devices. As the disease progresses, one must always remember to provide input.
2. Due to the progressive visual loss that the child experiences, professionals will need to direct their input to the auditory, tactile and motor modalities. Using these modalities, it is imperative that the teacher keep the child actively engaged in the activity being addressed.
3. Gain the child's attention before giving oral information (i.e., "Show me you are listening. Turn on your ears."). In addition, addressing the child by name will also gain his or her attention.
4. Listening is a passive activity. Incorporate the use of objects to elicit speech when you want the child to retell stories, answer questions, recite poems, and sequence rote information. When using this technique, the child's output of information is usually much greater in length and more intelligible. Active engagement is critical!
5. Begin early on to collect objects/toys that can be used in your lessons with the student. These concrete objects are used to actively engage the child in activities that enhance receptive and expressive language skills.
6. Allow the child to choose an object. Then, ask the child to tell all he or she can about the object. Encourage the child to analyze the size, texture, shape, composition, etc. This process of analyzing will enhance the forming of picture referents in the mind even when the child is not able to verbally express them.
7. Continue to read aloud from books, magazines, and newspapers. Engage the child by asking questions about what was read or what will happen next. Then, when answering

questions becomes too frustrating, provide two choices as possible answers. Have the student select the best answer. Yes/no questions can also be used.

8. If the child experiences difficulty with yes/no questions, the use of 'silly questions' is a good way to promote the processing of auditory information. For example, ask, "Do chairs eat?" "Do bananas talk?" Laugh about it and explain why it is silly. Have fun with these 'silly questions.'
9. 'Audio-describe' TV shows and activities that are taking place in the classroom and at home. Talk about what you see, hear, smell, touch and taste.
10. Tape record sound clues from field trips, vacations, home, school, etc., that can be used as prompts to aid in the recall of information.
11. Keep a taped journal or log of experiences and events that the student can listen to throughout the years.
12. Make a 'memory book' of favorite experiences and events written as stories along with a tactile representation cue attached to each page.
13. Over time, the facial and palatial muscles lose their mobility and preciseness. Exercise the facial muscles to enhance the intelligibility of speech by singing songs, reciting poems, and telling stories that use repetitive sounds and words.
14. Try singing a question to the student. Encourage the student to sing the answer.
15. A stress free environment will promote more intelligible speech.
16. Connected speech will be the least intelligible. Encourage two to three word utterances for more intelligible speech.
17. Combine speech work with physical therapy and occupational therapy activities.
18. Keep activities as 'hands-on' as possible by using objects, role playing, interviews, skits.
19. Continue to provide 'input' even when the child is not able to provide verbal 'output.'

Communication

Speech becomes difficult for the child partly due to muscle rigidity. Other factors include “deterioration of other cerebral functions such as memory and sense of grammar” (Hofman, p. 58). As speech worsens it can become difficult to be understood. One common problem is perseverations, where the child simply cannot break free from the speech pattern. Occasionally having the child sing song words will help as singing requires a different thought/muscle process than speech. The inability to find or articulate the right words is devastating to the child that has something to communicate. Add to this the ability to understand that far outweighs the ability to speak. Eventually the volume of speech declines and then will be lost completely. Alternative communications need to be considered long before this total loss happens.

Communication is the most important area of instruction for the student with Batten disease. Always teach with the long-term goal of enabling the student to communicate in the final stages of the disease. This is important to remember considering the child with Batten disease retains long-term memory more or less intact (Hofman, p. 68). As stated by von der Dunk and de Jong, “It is easy to overlook the fact that the ability to gauge the atmosphere can exist right up to the end. The understanding of the language also remains at a level which, compared to the limited and fragmental self-expression, is extremely high” (p. 22). Alternative ways to communicate will empower the student and give great comfort to the student in a time when many people underestimate the child’s ability to understand and know his or her own wants and needs.

Finding an alternative

First, if your student still has verbal ability, tape record his or her voice expressing favorite sayings, requesting favorite objects and activities, greetings, and anything else important to the student. Establish an audio-taped library of these expressions for use in the future. For instance, many augmentative communication devices are programmed with a real voice. Such devices, when they become necessary, can then be programmed with the student’s own voice. Hearing his or her own voice when verbal skills are diminishing is a source of comfort and empowerment and motivates the student to access this mode of communication. Often, the device will enable the student to access residual verbal ability by reducing the frustration of not being able to articulate appropriately without it. If speech is already gone, use friends' and/or family members' voices to program the augmentative communication device.

As the disease progresses, the child may not be able to make ‘voluntary’ movements to activate an augmentative communication device. When this occurs, it is still important to provide a way for the child to communicate. Try a simple augmentative communication device (2-4 locations) programmed with some of the child’s favorite sayings. Then, as the child accesses the device with involuntary movements, the device speaks and the child feels like he or she is still active in the environment. As the teacher, respond by saying something like “Good talking, Kevin,” “You have a lot to say today,” or “I like it when you talk to me.”

Next, pair activities with objects that represent the corresponding activities. For example, a young girl loves to ride horses. Her mother paired this activity with a rein. Then, every time the girl was going to ride horses she would hold this rein on her way to this favorite activity. Now, consider Jake, an older boy that does not know his prognosis, and how to introduce objects that represent activities in his school day. Following is his daily schedule and objects that the teacher cleverly introduced:

<u>Activity</u>	<u>Object Representing the Activity</u> (underlined)
Calendar Time	Jake arrives at school and removes his <u>absentee card</u> from its holder to turn in to the teacher at the appropriate time.
Orientation and Mobility	Jake's job is to practice his orientation and mobility by taking the <u>attendance cards</u> to the office.
Reading	Jake listens to a book on <u>cassette tape</u> while following along in his Braille reader.
Recess	Jake loves to jump rope at recess. He carries his own <u>jump rope</u> out to recess.
Math	Jake uses a <u>talking calculator</u> to complete his math problems.
Lunch	Jake carries his <u>lunch card</u> to the cafeteria.
Physical Therapy	Again, Jake practices his orientation and mobility while building stamina by walking to the faculty room where he buys a pop with coins from his <u>coin purse</u> .
Music Therapy	Jake's job is to start music class by ringing the <u>bell</u> .
Bus	Jake carries his <u>notebook</u> home with a note telling his family about his day.

Once the objects are clearly established in the student's mind, switch to a part of the object, when possible, to represent the activity. For example, the girl who loves to ride horses begins to associate the activity with a piece of rein rather than the whole rein. These parts of objects can then be attached with Velcro to an augmentative device.

In this way, the student with visual impairment and limited verbal abilities can find the part of the object and press it to activate the augmentative device to request the activity (Fig. 2). Later, when the ability to differentiate the parts of objects becomes limited, return to the whole object to allow the student to continue to request desired activities, wants, and needs (Fig. 3). Objects and parts of objects are more concrete than textures or Braille and are easier for the student to continue to distinguish even when abilities to differentiate become limited. Also, teaching the objects early on in the process helps to establish the concept in the student's long-term memory. Again, this is important because long-term memory appears stronger than short-term memory as the disease progresses.

Figure 2: Augmentative device for expressive communication.

Figure 3: Augmentative with objects attached for tactile recognition.

Calendar systems or schedules

The student with Batten disease may feel out of control as he or she experiences the regression of skills and the reduced ability to express oneself adequately. The student may communicate this feeling through restlessness, irritability, withdrawal, aggression, or changes of mood (Hofman, p. 33). Providing structure in this student's day will assist the student feel more in control. Utilizing a calendar system for the student with Batten disease is an excellent way to both provide structure and to practice the communication system of objects/parts of objects corresponding to activities.

According to Downing and Peckham-Hardin, "Students can use schedules to request a specific activity, ask when something will occur, or to answer direct questions regarding their day" (p. 62). In this way, students practice their communication skills. Other skills can also be practiced while utilizing calendar systems or schedules. For example, a series of boxes that close allows the student to practice fine motor skills when opening each box, and a stationary schedule allows the student to practice mobility while walking to the schedule between each activity. Reading, writing, and math can also be practiced in the context of the schedule by reading the Braille word that accompanies the object, writing the day's schedule in the morning by referring to the schedule, and practicing counting and sequencing while manipulating the calendar system (Downing and Peckham-Hardin, p. 67).

As the disease progresses, the calendar system or schedule will provide the necessary structure to reduce the chaos that is happening to the student. Emily, for instance, began with a system consisting of a series of eight boxes. Each box contained an object that corresponded to an activity planned for that day. Assisted by her teacher, Emily would open the first box, complete the activity, return the object to a 'finished' box, and repeat the process with the next box. A great deal of interaction happened between Emily and her teacher throughout this process. The number of boxes in the system decreased as time went on. However, the structure of the calendar system brought comfort to Emily. She knew what was going to happen in her day. When Emily became agitated, merely holding the calendar box would calm her and the agitation would vanish.

Calendar systems or schedules can take on a variety of appearances depending on the needs and desires of the child. A few ideas include pictorial schedules, tactile schedules, systems in a book format or as part of a school notebook, Velcroed symbols on a clipboard, symbols in a slide holder, and objects in a series of boxes (Downing and Peckham-Hardin, p. 62-63). One teacher uses a shoe holder that hangs on a door to place objects in that corresponds to the activities of the day. Very creatively, one mother even developed an auditory system of music to prompt her child to the corresponding activity. The key to a calendar system or schedule is not the appearance but the consistency of its use.

Daily calendars

A routine or pattern is established when the calendar box objects are used in a set order time after time. This is recommended when beginning a calendar system to enable the student to mentally connect the object with the activity. When setting up a calendar system, it is recommended to run it consistently for a one month period before making any changes. This allows sufficient time to establish understanding of the objects or symbols and of the routine. At the end of one month,

make any necessary changes and run for another month. Continue in this fashion until the student understands the function of the calendar box.

The next step in calendaring is to add objects or symbols that represent additional activities. The student will soon have more objects/activities than can be accomplished in one day. At this time, the student begins to make choices by choosing what activities he or she wants and in what order. Choices are made at the beginning of the day, so the student is actually planning his or her own day. As choices are made, the object or symbol goes into the box at the possible position. This choosing may take place in only one position of the calendar box to begin with, but the choice empowers the student, in any case. The teacher still maintains whatever control is necessary by offering only choices that are possible for that day and only in the box position that allows for choices.

Example: Upon arrival to school, Emily is given six of her calendar box objects. The teacher then asks Emily what she wants to do first. Emily loves lunch, so she chooses the spoon that represents lunch. The teacher says, "Good choosing, Emily, but lunch isn't until 12:00. Let's put the spoon in box #3. What else would you like to do today?" This process continues until all four of Emily's boxes are filled. The teacher then says, "You planned a very good day. Let's get started. Please find box #1."

In the example, the teacher maintained the control she needed while still allowing Emily to make choices. Behavior problems are quickly reduced when empowering the student with some control over his or her day. Remember, though, the teacher is still in control over the offered choices.

Sometimes it is not possible to offer the student a choice. For instance, the physical therapist may be scheduled to come in and do range of motion with the student right before lunch. In this case, the teacher may present the student with two objects and ask which object represents range of motion. Again, the student is actively engaged in the planning of his or her day.

One type of calendar box system that works well consists of two magazine holders attached together back-to-back (Fig. 4). One side will hold up to five small boxes that are the basis of the calendar system. Each box will have a number or dots on it designating its position. Use raised numbers, dots, or Braille numbers when using the system with a visually impaired student. The other side of this system is called the 'finished box.' Upon completion of an activity, the student places the corresponding object or symbol in the "finished box." This is an excellent way to identify the close of an activity.

A great deal of verbal communication between teacher and student must take place while interacting with the calendar box. In fact, the presence of the calendar box actually reminds the teacher to communicate with the student. All too often, children with disabilities have things done to them or for them, rather than with them. Co-active involvement with the calendar box will reduce the occurrence of this problem. Again, the student feels empowered when the calendar box is used appropriately.

Figure 4: Calendar box system with 'finished' side.

Calendar box hints

Following are some useful hints to remember when implementing calendar boxes. Note the difference between the acquisition phase and the application phase. Also remember, interaction with the student throughout the calendaring experience will increase the likelihood of success. We must assume that the student understands everything we say. Include academic concepts within your conversations. Who knows? You may open up a whole new world for that very important student of yours!

Acquisition Phase:

1. Start small. Pick a time in the student's daily schedule that occurs consistently. Start the calendaring system there, even if you start with only two boxes. Consistency is the key at the acquisition phase.
2. Implement the system every day. At least begin the schedule. If the day falls apart, discontinue the routine and start again tomorrow. Order is very important in this phase.
3. Never skip a box. Complete the schedule in order. If time is short, do the corresponding activity briefly. Order and consistency will enhance the student's learning at this phase.
4. Attach a written schedule directly to the boxes. Run this schedule for one month. Avoid the urge to make changes until this time. Then, make the necessary changes, post the new written schedule, and run the system for another month. Repeat this process as often as is necessary. Changes may include changing the order or adding another box.
5. Do not give up. Every student can learn. Some students just need more practice and consistency. Failure to succeed at calendaring is most often attributed to the caregiver giving up before the student has had ample opportunity to acquire the skill. The process may seem long but is so worthwhile to the student who may have little control over his or her life.
6. Observe, observe, observe. Look for those small cues that signify the student has acquired the knowledge that the object represents a certain activity. This could take days, weeks, or even months! Keep going, though. It is that important. Build those small cues to more definite ones as the skill becomes ingrained.

Application Phase:

1. Empower the student by allowing him or her to help plan part of the schedule. Place two object choices in one box that comes at a point in the schedule that a choice of activities is possible. Allow the student to choose between the two objects. Accept even the slightest movement toward a choice. Honor the student's choice and reinforce the choosing.
2. Some students may reach the level of ability that they can actually plan their own day. What power you have given a student that may have everything done and said for him or her at this point. You have truly increased this student's quality of life.

Early experience with objects

Early experience with objects is critical to the facilitation of communication when verbal ability is difficult to access or gone altogether. In fact, the earlier the better, due to the retention of long-term memory.

If your student is still verbal and not requiring adaptive communication, you still need to provide experiences with objects. Accomplish this by adding objects as often as possible to the curriculum you are teaching. For example, use actual objects when teaching math concepts. Allow the student to use manipulatives when practicing counting, addition, subtraction, etc. Teach spelling words that can be represented by the object that you present. Use this same tactic for practicing speech by having the student say the word that represents the object.

Games are always a fun way to provide experience with objects. Try presenting a secret sack with objects inside. Students take turns reaching into the sack and guessing what the object is that they choose without looking. Playing Bingo is another fun way to experience objects. The teacher presents an object and the students find the word on their bingo card that corresponds to the object. Try reversing this by providing bingo cards with small objects attached. The student then finds the square that corresponds to the word that the teacher reads or writes on the chalkboard. Be creative and have fun with this!

Sorting and classifying are two powerful activities that provide experience with objects. Both activities provide the student with the knowledge that objects can vary yet still be classified as the same. For instance, a comb is a comb whether it has large teeth or small teeth. Also, a comb may have a handle or not and still be a comb. The ability to classify and sort may solve problems down the road. For example, a student may use a favorite spoon to represent lunch. However, if the particular spoon that is used gets lost the student will more easily switch to a different spoon to represent lunch even if the spoon appears very different.

Finally, use objects as cues or prompts to activities whenever possible. One example is a young boy, Cody, who loves to go to grandma's house. His parents let Cody carry the keys to the car every time that the family is going to visit grandma. The same boy wears a small wristwatch when traveling to school on the bus. Another example is to collect small souvenirs of trips, visits, weddings, and so on (Johnston, p. 4) to reference when the child wants to talk about the occasions. These objects will be helpful in determining what the child wants to talk about when he or she is no longer able to express his desires. Then, the child can present the caregiver with the object to signify the topic to discuss. In this way, frustration and agitation can be avoided when the child has no other way to express himself. Refer to the previous section on finding an alternative for more examples. Remember to always reference the objects when they are being correlated with an activity.

As changes occur in classrooms, classmates, and teachers, the objects that represent the student's communication system stay consistent. Predictable objects/activities can help to make a new environment less strange (Johnston, p. 4), enabling smoother transitions and less frustration. According to von der Dunk and de Jong, "Precisely because the world becomes more confusing and unpredictable, due to the progressive loss of their grip on the order of things, they develop a

need for stability, which can be satisfied by looking back to the past, to what they know” (p. 42). Consequently, provide early experience with objects. Let objects become what the student knows and relies on for future communication purposes. The time working with objects will likely be the best time spent.

Experience books

The student’s understanding of language supports the need to be talked to, sung to, and read to throughout the disease (Schrek, 2002). Von der Dunk and de Jong (1992) recommend that this communication be more than just talking to the children. Rather, the communicator must try to understand what the child is experiencing and communicate accordingly, remembering that the child understands at quite a normal level. Experience books will allow the communicator to have a reference of what is important to the child and what he or she may want to talk about. Also, children with Batten disease love to reminisce about the past.

An experience book is a book about the individual student. Each page tells a story about an experience that the child has encountered. Each page also has a tactile representation of the activity that the child experienced (Lewis & Tolla, 2003; Monaco & Mamer, 1993). These are collected from family events, field trips, and school activities, and used to tell the child’s story as a point of reference in the future (Johnston, 2001; von der Dunk & de Jong, 1992). A student that knows Braille may write his or her own story. A student that still has verbal ability may tell his or her own story for the teacher to write or Braille. Regardless of how the experience book is produced it will provide a rich source of memories and be an aid to conversation between the student and the teacher, peer, family member, or any other communicative partner (Johnston, 2001). Students often want to talk about past experiences, even when their verbal ability is diminished and hard to decipher. The student is able to locate the object on the page that he or she wants to discuss allowing others to carry on a meaningful conversation with the child with the information that is written. Thus, experience books are motivating and a source of pride for the student. Some students even have their books recorded on cassette tapes for listening to in the future when motor skills have diminished and the books can no longer be accessed independently.

Summary of Best Practice

Calendar Systems: Calendar systems provide structure for the students and predictable routines throughout the day. The object cues within the calendar system provide the students with concrete representations that are easily distinguished and recognizable. The object cues also provide the students with an acceptable item to fidget with during activities thus satisfying this sensory need common to students with Batten disease.

Choices: Making choices in one's life is empowering. This is an important aspect to consider when educating students with Batten disease. So much in their lives is being dictated by the disease taking away skills they once had. The students are keenly aware of this painful fact. Thus, making choices about activities and within activities provides the students with some control.

Experience Books: Experience books or scrapbooks are utilized as a positive behavior support strategy. This strategy allows the student with Batten disease to reminisce about past experiences. The experience book with tactile additions allows the student to initiate conversation about topics he or she enjoyed by choosing the page with the story he or she wants to discuss. The stories allow the conversation partner to know exactly what the student wants to discuss. Thus, the frustration level of the student is likely to decrease because the topic is clearly understood by the conversation partner.

Music: Music can be used to comfort the student during times of agitation and to help redirect them back to a state of calmness, which allows him or her to continue the day productively.

Augmentative Devices: All students with Batten disease eventually lose their ability to speak. Augmentative devices can be utilized in the form of multiple-location switches and one-location pressure switches. Adding a texture or object to the switch locations allows the students to understand which location would 'say' the phrase they wanted to communicate. Also, the application of a one-location pressure switch to activate a book-on-tape or music allows the students to be active learners in the activities rather than just passively listening. The active participation of maintaining pressure on the switch appears to help the students attend to the task for a longer period of time than they would if just listening.

Consider the use of series books for all age groups. The little ones may enjoy Dora, Sesame Street, and Disney books. Older students may enjoy American Girl, the Vampire series, and the Box Car Children. The familiarity of the characters will add to the enjoyment of listening to these books on tape. A special object could represent the whole series.

Redirection: Redirection from behaving inappropriately is a strategy that can be used as a positive behavior support strategy. Redirection occurs through a number of ways: Providing an object cue for another activity, providing choices, providing access to the experience book, and counting, singing, or telling a story during a less preferred activity.

Honoring Choices or Requests: Providing choices is only effective if those choices are honored. For example, the choice of when to participate in an activity or allowing the student to dictate when the activity ends empowers the student if the choice and/or request is honored. This strategy can help decrease inappropriate behaviors.

Individualized Education Plan (IEP) Goals

IEP goals can be written to directly involve the calendar box. Following are examples of IEP goals used to increase skills in basic communication and in muscular control.

Basic Communication:

1. When presented with an activity/object, _____ will vocalize to show anticipation of the routine/activity 80% of the time over three consecutive data days.
2. Throughout the day when _____ is presented with his calendar box, he will reach out and touch the object to indicate his desire to do the corresponding activity 75% of the time for three consecutive data days.
3. Throughout the day when _____ is presented with her calendar box, she will reach and grasp the object to indicate her desire to do the corresponding activity 80% of the time for three consecutive data days.
4. When presented with an object, _____ will Braille the activity that corresponds with that object and spell the words with 90% accuracy over three consecutive data sessions.
5. When presented with a variety of calendar box objects, _____ will place the objects in the correct order on his schedule at 100% accuracy as the teacher reports the upcoming day's activities for a consecutive five days.

Muscular Control:

1. When presented with his calendar box and given verbal prompts, _____ will open his calendar boxes at 75% accuracy over three consecutive data days.
2. When presented with a calendar box to _____'s left side and paired with an auditory cue, _____ will actively turn her head towards the calendar box (moving from right to left) for 75% of trials over three consecutive data days.
3. When presented with a calendar box object, _____ will use his cane to trail the wall and locate the area where the activity will take place with no verbal prompts to two different locations for three consecutive data days.

As you can see, calendar box goals can be directly related to other goal areas needed by children with disabilities. The calendar box provides motivation to work on tiresome goals by empowering the student with choices. This may be the only empowerment that some students have. Also, the calendar box enables the teacher to provide organization and the stability of a routine to the life of his or her students.

Making choices is important to all students. Making choices is no less important to the student with Batten disease. When verbal skills have declined, the student with Batten disease can continue to make choices by choosing from the objects that correspond with activities. Even in the final stage of the disease, the student that has a communication system of objects can still make choices. This is accomplished by placing each of the student's hands on an object. Carefully observe for any movement, even a slight movement of the finger. This movement will designate the student's choice. What empowerment this choice-making ability will give the student!

Educational Strategies for Children with Infantile and Late Infantile Batten Disease

A child who has little ability to interact, both physically and vocally, may lose motivation if subtle attempts at interaction are repeatedly missed. An example of this may be a child who can only make soft sounds when uncomfortable will stop trying to communicate this need to move if the subtle sounds go unnoticed. The child may think, “What’s the point? No one can hear me anyway. I may as well just go to sleep.” Thus, this child becomes a passive being in a world that moves too quickly to notice. To motivate and engage the passive child requires an observant, caring teacher. Following are strategies aimed at teaching the child to interact with his or her environment to the extent possible.

Establish a ‘yes/no’

A child that has a clear ‘Yes/No’ communication is very empowered to interact with others, even if that is the only communication possible. The child with a ‘yes/no’ is able to answer questions concerning wants, needs, comfort, and feelings. The child with Batten disease is able to cognitively know these things, and a ‘yes/no’ communication is not only empowering but comforting.

The teacher must carefully observe the child who has little physical and vocal abilities. Look for what the child **CAN** do. For instance, Hanna had no controlled movement of her legs, arms, or head. In fact, the only function she appeared to control was her eyes, which were very animated. The teacher observed this and chose to build on this strength. The teacher placed Hanna on her lap while sitting in a rocking chair. Back and forth. Back and forth. Hanna enjoyed the rocking very much. Upon stopping, the teacher asked Hanna if she wanted ‘More.’ The natural movement of the rocking chair and subsequent stopping naturally helped Hanna’s eyes move up. The teacher then responded that Hanna asked for ‘more’ through responding ‘yes’ by moving her eyes up. Within days, Hanna knew that if she moved her eyes up she was responding positively to yes/no questions. This opened up a whole new world for Hanna. She could now respond to yes/no questions concerning positions she wanted to be in, if she wanted certain music, and many other questions. How empowering! Over time, Hanna became much more engaged with her environment, laughing at jokes and making verbal noises. Hanna even learned to ask for music by making a certain noise using her tongue and voice.

Turn-taking

Turn-taking is a great activity that encourages communication between the student and the adult. Begin by imitating what the child does. Wait. Then reinforce if the child repeats the imitation. An example of this is Dion. Dion was thought to be non-communicative. However, turn-taking proved this to be wrong. Dion was able to make a small squeak noise that was often accompanied with a smile. Dion’s new teacher knew about turn-taking and tried this strategy with Dion. When Dion squeaked, the teacher followed with a squeak. She then waited. Dion stopped all sound and

movement. The look on his face told the teacher that he was processing this new interaction. After about ten seconds, Dion broke into a smile and squeaked even louder than the first time. What great fun the teacher and Dion had taking turns and conversing in the language Dion could master.

In just one session, Dion learned that he could also ask for 'More' by making this same squeak. Children with Batten disease are bright. Never underestimate their ability to communicate. The key, however, to successful communication is through the adults in the environment. Observe, respond, interact, and reinforce. You, as the teacher, have the power to open the world of communication for your special student with Batten disease.

The 'Little Room'

Danish teacher-therapist Lilli Nielsen developed the strategy of the 'Little Room' for children with visual impairments who were extremely passive (Dunnnett, 1997). A 'Little Room' is a plexiglass enclosure consisting of a top and three sides. The plexiglass has holes throughout for ventilation and to allow for objects to be attached. A variety of objects are hung on the sides and from the top, attached with elastic (Fig. 5). Objects used may include a brush, beads, cup, ball, plastic keys, bell, spoon, hair curler, spring, hair pick, soap drainer, mesh bag full of marbles, toys, sound makers, etc. A variety of textures may also be attached to the sides or top (i.e., mylar, waxed paper, bubble-wrap, metallic tissue paper, etc.). Do not move the position of the objects/textures.

Place the 'Little Room' over the child in the position most favored. Allow the hanging objects/textures to hang close to the child's face and hands. Allow the child to spend time inside the 'Little Room.' Eventually, the child will make a voluntary or involuntary movement and bump one of the objects or textures. As time progresses, the child will voluntarily or involuntarily continue to make contact with the objects/textures in the 'Little Room.' The movements that interact with the objects/textures result in the child realizing that there is something "out there" (Dunnnett, 1997). The goal is for the accidental movement to lead to conscious exploring of the environment. By keeping all items in the same position the child becomes confident in reaching out and knowing what to expect.

Implement the 'Little Room' in an area as quiet as possible so as not to distract the child. Also, do not interrupt the child's independent exploration by verbally reinforcing or prompting to repeat an interaction. The child needs quiet concentration while in the 'Little Room.' Likewise, the teacher should engage in quiet observation of the child to understand the level of cognitive interaction exhibited.

Hopefully, use of the 'Little Room' will encourage the child with Batten disease to continue to interact with the environment for as long as possible. The child no longer able to physically interact may enjoy the 'Little Room' with visual stimulation attached to the sides and top.

Figure 5: Lilli Nielsen's 'Little Room.' Frame is made from $\frac{3}{4}$ " PVC pipe. Dimensions vary – big or small as needed. Objects are any manipulatives the child enjoys.

Resonance board

Resonance boards are hollow platforms that allow the user to perceive sounds through vibrations. This intervention strategy was originally developed by Lilli Nielsen for use with children with visual impairments and blindness (Brown).

You will need a square piece of plywood at least 1/8 inch thick to construct a resonance board. A heavier child may need thicker plywood. Four feet square is a good size but can be adjusted according to the size of the child.

Next, attach four strips of 1" X 1" wood to the underside rim of the plywood (Fig. 6). Finishing the plywood is important as you want a very smooth surface for the child to lie on. Use sandpaper and then glasspaper for this process. Once the plywood is very smooth, apply two coats of furniture wax with a polishing cloth. The work to smooth the resonance board is worth the reaction you'll get from the child.

If the child is extremely sensitive, try laying a towel on the resonance board before placing the child on it. This will muffle the sensation a bit. Another strategy is to lay the child on carpet next to the resonance board with only his or her feet resting on top of the board. Any movement the child makes on the surface of the resonance board will result in amplified sound and matching vibration. Therefore, even a small movement will cause an effect.

Experiment with using a variety of objects on the resonance board. Begin by providing the interaction necessary to provide the child with the level of stimulation that is tolerated. This may be dragging a dog chain across the surface, tapping different locations on the board with various items, or activating a vibrating or wind-up toy on the board. Watch the child's reactions to each.

Try placing the 'Little Room' over the child while he or she is lying on the resonance board. This combination of strategies enhances any movement the child makes and provides feedback and reinforcement for moving. Also, encourage turn-taking with the child while on the resonance board.

Interestingly, a distractible, active child often will remain on the resonance board to receive the feedback available there. On the reverse, a very passive child is likely to become more active due to the ease in which he or she is able to produce sound and vibration. Also, a vocal child may become silent in order to listen, but a silent child may vocalize for the resultant sounds and vibration. In short, "children who do not normally interact with other people in positive ways may attend, wait, look, touch" (Brown, p. 4) during turn-taking activities while on the resonance board.

Calendar boxes

The calendaring process is the same for children with infantile and late infantile but at a much simpler level. For instance, you can still place an object that corresponds with an activity in their hand each time. You might even try different bracelets and/or necklaces that they can wear if holding objects is a problem. Another strategy is to have the child wear a bib with Velcro attached to allow an object to stay with the child throughout the activity.

Figure 6: Lilli Nielsen's resonance board. Plywood veneer attached to a 1" x 1" frame. Plywood veneer sized to accommodate individual – child should lie comfortably on the veneer. Plywood veneer needs to be extremely smooth.

The main difference is the extent of experiences that the little ones have had. However, there will still be activities that they respond to and enjoy – going in the car, visiting grandma, listening to music, rocking with mom, going for a walk with dad, etc. Daily care activities should also have an object such as a diaper for changing time, a bath mitt for bath time, a syringe for tube feeding, etc. Lotion is a fun object to represent getting dressed. Help the child rub the lotion in using his or her hand, even if the activity is totally hand-over-hand. In this way, the child gets input where the lotion is rubbed as well as on the palm of the hand that is rubbing. Help the child touch and/or hold the object each time the activity happens. Bring both hands to midline and have the child feel the object with both hands as often as possible. Talk about the object and what is about to happen. You may want to use simpler language but *never* baby talk. Remember, these children are in there but locked in a body that prevents full interaction.

Calendar boxes provide the forum for adult interaction that will support the child emotionally (Blaha and Moss, 1997). Enjoy this time with your special student.

Augmentative devices

Any augmentative device will work if it can be accessed with a light touch. Also, the device needs Velcro attached. Therefore, you may consider a device that is fairly reasonably priced. An example is the Cheap Talk. It is also very easy and fast to program. The four-location device that is horizontally placed works well as it fits on a wheelchair tray and all locations are within close proximity to the child. Attach the object to the corresponding message on the device with Velcro. You may need to help the child access the device using a hand-over-hand technique. Nevertheless, the child will be participating in a meaningful way.

There are also a number of reasonably priced pressure switches that works well to engage the children in activities. For instance, attach a radio, tape player, or any battery operated toy to the one-location pressure switch. Finally, place the switch under the child's hand, arm, foot, etc. to allow the child to keep pressure on the switch. In this way, the child is an active rather than passive participant in the activity.

Experience book

The Experience Book works the same way for all children with Batten disease. However, you will have to provide the book to the children with Infantile and Late Infantile Batten Disease. The Experience Book is motivational for the child because it is about him or her. Remember to make the book in a fashion that relates the child's experiences (Monaco and Mamer, 1993). Like the calendar box, help the child touch the tactile representations with both hands as much as possible. Ask questions about the story and watch for a yes/no response to engage the child in the activity.

Behavior Management

Behavior management is an area that often must be addressed at some point or another with a student with Batten disease. *Therefore, it is of utmost importance for the teacher to fully understand what the child is experiencing as the disease progresses so as to apply the appropriate behavior intervention strategies.* However, von der Dunk and de Jong (1992) caution that “understanding is not synonymous with excusing everything and expecting nothing” (p. 49). Von der Dunk and de Jong (1992) also report that some characteristics that the child had before the onset of the disease may present as more pronounced at a later stage of the disease. For example, a very independent young child may try to be even more independent in later stages regardless of the increasing dependence on others that results from the disease process. As might be expected, this scenario may cause great frustration to the child and may lead to acting out behaviors. Personality and characteristics of the child also influence the agitation level experienced and displayed by the individual child.

Behaviors related to the disease

The first thing to think about when dealing with inappropriate behaviors is the fact that ability levels of the student are declining. Teachers may often interpret task-avoidance behaviors as non-compliance when actually the inappropriate behaviors may be due to frustration over the inability to complete previously mastered tasks and the increasingly limited control over performance (Koehler and Loftin, 1998). Perhaps it is easier to avoid the task than to face certain failure. Koehler and Loftin (1998) report, “As it becomes increasingly difficult for the children to perform more and more tasks, they may be prone to more frequent outbursts of anger, episodes of crying, and the like” (p. 323). These outbursts may actually point to a deeper feeling of stress than the immediate cause of the outburst would justify (von der Dunk and de Jong, 1992).

Koehler and Loftin (1998) report:

Acting-out behaviors also seem to be more likely at those stages when significant changes become apparent that affect the child’s day-to-day life, such as a shift from reading print to reading Braille, the use of a cane for mobility, the use of a wheelchair or walker, and the beginning of seizures. The onset of adolescence is also a time when such behaviors frequently emerge” (p. 324).

However, the student is usually frightened and angry over the immediate change rather than the long-range prognosis of the disease (Koehler and Loftin, 1998). Along with fear and anger, sensitivity to failure, frustration, emotional disturbance, anxieties, and aggression are other common emotional symptoms experienced due to the disease process (Juvenile Batten’s Disease, 1999). These emotional experiences may begin as early as during the initial phases of deterioration and are still present during the final phase of the disease (Hofman, 1998). When dealing with behavior problems, the teacher must be aware that the students suffer from the disease process and it frightens, shocks, and confuses them (von der Dunk and de Jong, 1992). In addition, Hofman (1998) reports that the children were aware of or had a feeling about the decline that is happening for a very long time. As a result, the teacher that will be most successful with these special students

is a sensitive teacher. One that will talk to them, not about them, and will instill a feeling that they will not be left on their own. This teacher will help with whatever happens, with whatever burdens the children will have to bear (von der Dunk and de Jong, 1992). Some teachers feel this is a monumental task that they are not prepared to accept. However, the rewards of helping the child and the family through a struggle that they did not ask for far outweigh the hardships.

Besides the decline in ability and the upset of emotional experiences, students with Batten disease also experience delusions and hallucinations as the brain deteriorates (Juvenile Batten's Disease, 1999). Behaviors presented may include hysterical laughing or crying. The hallucinations appear to seem extremely real to the children and can be very frightening. Hofman (1998) also recommends a calm, consistent approach by staff in a dependable and supportive environment to help bring the child back to reality. Von der Dunk and de Jong (1992) add:

Expressions of acute psychoses also occur. It is important to be able to identify these as such and to take into account that the *behavior* is probably not caused by a reaction to circumstances, but has an organic source. This means that staff must be present, but that it will not help if they try to regulate the behaviour by manipulating the environment. The most effective solution is to let it happen, in the knowledge that it will pass" (p. 40).

Therein lies the challenge – determining the cause of the problem behaviors. Are the behaviors a symptom of the disease or typical problem behaviors expressed by other students? Determining the cause is important when designing a behavior management plan for the student with Batten disease. Koehler and Loftin (1994) state, "Maintaining a realistic level of expectations for the child may be one of the most difficult tasks that a teacher faces" (p. 323). Are our expectations for the child maintaining or increasing problem behaviors? Knowledge of the child and the disease and an assessment of the environment will help answer this question.

Assess the environment

Daniels (1998) offers eight questions to ask yourself, as the teacher, when analyzing the cause of a student's problem behavior. This process is a great place to start prior to developing a behavior management plan. Let's look at each question and relate it to the student with Batten disease.

1. *"Could this misbehavior be a result of inappropriate curriculum or teaching strategies?"* (p. 26).

Students with Batten disease retain long-term memories of what they could do prior to the deterioration of the disease. They still want to belong, to be a part of things. Include them in all teaching experiences by talking to them, explaining the meaning of the curriculum, and asking questions in such a way that they can respond without too much effort (von der Dunk and de Jong, 1992). Daniels (1998) also recommends making the curriculum meaningful to the student by first analyzing the futuristic benefit for the child.

2. *“Could this misbehavior be a result of the student’s inability to understand the concepts being taught?”* (p. 26).

Again, Daniels (1998) recommends the demonstration of how the skills being taught have meaning to the student. This will aid understanding. However, the inability to understand the concepts being taught is often not the reason for misbehavior in children with Batten disease. Rather, they are often treated too childish and talked down to which only aggravates them further. Although they are wrestling with deterioration and feelings of helplessness, these students respond to and deserve age-appropriate concepts to be taught. Too often their school activities become less meaningful and *they know it* (Hofman, 1998). The trick is to balance a meaningful curriculum with appropriate adaptations to allow the student to participate.

3. *“Could this misbehavior be an underlying result of the student’s disability?”* (p. 27).

Misbehaviors due to the student’s disability are highly likely, especially if the student rarely or never displayed inappropriate behaviors prior to the deterioration caused by the disease. A review of the section on behaviors related to the disease will be helpful when planning, changing, or implementing any behavior management program. Daniels (1998) recommends being flexible. Avoid strict adherence to old teaching strategies that may be unsuccessful and inappropriate when teaching the student with Batten disease.

4. *“Could this misbehavior be a result of other factors?”* (p. 28).

Daniels (1998) suggests the following factors that may cause misbehavior: 1) the physical arrangement of the classroom, 2) boredom or frustration, and 3) transitional periods. For the child with Batten disease and visual impairment or blindness, the physical arrangement of the classroom is important. Explain and demonstrate where specific areas are in the classroom. Be sure that pathways between areas are free of obstacles. It is helpful to maintain the same classroom arrangement for the entire school year for a feeling of safety by any student that is blind or visually impaired. In addition, the classroom should have an area that is designated for quiet time or relaxation. This area can become a haven for the student that becomes overwhelmed with the activity or the feelings that he or she happens to experience.

Structured schedules that alternate easy activities with more challenging activities and seatwork with physical activities will help the student through periods of boredom and frustration. The schedule must be predictable. In this way, the student is able to look forward to favored activities that may help him or her get through less preferred activities. Transitional periods are also less stressful when the student follows a predictable schedule. Reducing the level of chaos during transitions will also help the student. For example, release the student five or ten minutes early to travel to the next class, to recess, or to the bus when the halls are empty.

5. *“Are there causes of misbehavior that I can control?”* (p. 28).

Daniels (1998) specifies that the teacher is in control of modifying or changing the curriculum, adapting instruction, changing his or her communication style, changing his or her attitude and expectations of the student, and giving more positive feedback. Von der Dunk and de Jong (1992) recommend adopting an attitude that allows the child with Batten disease to enjoy life. They go on to state, “What that child experiences now can mean a lot later on, because it may no longer be possible at a later stage” (p. 23).

6. *“How do I determine if the misbehavior is classroom based?”* (p. 28).

The best way to determine if the misbehavior is classroom based is to conduct a functional analysis to examine cause-effect relationships. Specific information concerning how to determine these cause-effect relationships follows in the section on functional behavioral assessment and analysis.

7. *“How do I use reinforcement strategies to reduce disruptive behaviors?”* (p.29).

Daniels (1998) reports that teachers deliver positive reinforcement through words, physical expressions, closeness, activities, and primary reinforcers. It is all too often the case that students are ignored or taken for granted when their behavior is good but addressed by the teacher when their behavior is bad. Remember that positive reinforcement should be delivered at a minimum rate of four per one negative comment. More is definitely better. Try aiming for eight positive comments, physical expressions (i.e., wink, thumbs up, etc.), closeness, activities, or primary reinforcers per one negative comment. Many times students want the attention of the teacher, and they do not particularly care if it is positive or negative. Turn your classroom into a positive environment and watch the students’ behavior improve.

8. *“Is it appropriate for me to use punishment?”* (p. 30).

Punishment must always be used with extreme caution. Punishment is effective to suppress an inappropriate behavior but does not guarantee that the student knows appropriate behavior to replace it (Maag, 2001). A behavior does not simply go away. The behavior will be replaced. If the teacher does not teach an appropriate replacement behavior, the student will replace it, sometimes with a worse behavior than the one being replaced. Maag (2001) states, “Teaching involves giving children skills and knowledge, not suppressing or eliminating behavior” (p. 178).

Functional behavioral assessment and analysis

A functional behavioral assessment includes interviews, direct observation, and an examination of school records (Condon and Tobin, 2001). O’Neill, Horner, Albin, Sprague, Storey, and Newton (1997) offer an interview form that is utilized with the team of teachers that work with the student. The interview is extensive and provides the observers with target behaviors to be analyzed following the direct observation. Setting events, events that may influence the likelihood of the inappropriate behaviors, are also discussed.

O'Neill et al. (1997) provide a functional assessment observation form for observers to record the time, the behaviors, the predictors (or antecedents), the perceived functions of the behaviors, and the actual consequences that follow the behaviors. Following the observation, the teacher examines the form for clusters or groupings of recordings that provide information about the behaviors, what sets them off, why the student exhibits the behaviors, and the consequences that either decrease or escalate misbehaviors.

Predictors, or antecedents, to the behavior may include demand/request, difficult task, transitions, interruptions, alone or no attention (O'Neill et al., 1997). The time of day may also be a predictor if behaviors cluster around certain time periods such as morning, just before lunch, after lunch, mid-afternoon, etc.

If a *demand/request* sets off a behavior, consider the time of day that such demands/requests are required. Also, consider behavior momentum, a strategy where the teacher encourages the student to comply by instigating several easy requests prior to a more difficult, less preferred demand. For instance, the chain of requests might be: 1) "Give me five!" 2) "Have a drink of milk" (a preference), and 3) "Take a bite of potatoes" (a less preferred task due to difficulty in swallowing).

If a *difficult task* sets off a behavior, again consider the time of day that the difficult task appears in the daily schedule. Also, consider adapting the task to allow for a higher likelihood of success and to maximize opportunities for independence (Koehler and Loftin, 1998). Remember, too, that high rates of positive reinforcement will increase the likelihood that difficult tasks will be attempted with less non-compliance.

If *transitions* set off behaviors, implement a calendar box system to allow the student to predict what will happen throughout the day. A token economy system (i.e., the student earns tokens for appropriate behavior and is allowed to spend them for a reinforcer when enough tokens are accumulated) is also likely to increase appropriate behaviors during transitions.

A calendar box system is likely to decrease misbehaviors during *interruptions* because the student learns to anticipate activities and to predict the continuation of an activity later in the schedule. A timer will also help. For example, tell the student that he or she can continue an activity for two more minutes, set the timer for two minutes, and then discontinue the activity when the timer goes off.

Misbehavior that occurs following periods of *alone (no attention)* may be addressed by using peer tutors to interact with the student when the teacher is unavailable. Also, check the level of work that is required during individual work time. Ensure that the work is at a level that the student can complete it successfully when working alone. In addition, be aware of the amount of unengaged time that occurs throughout a typical day. Boredom during unengaged time is likely to increase problem behaviors.

Perceived functions of misbehavior include to get/obtain something or to escape/avoid something. The get/obtain function of a behavior includes getting attention, a desired item or activity, self-stimulation, etc. The escape/avoid function of a behavior includes escaping a demand/request, an activity, a person, etc. Condon and Tobin (2001) recommend creating environments where the problem behavior is not necessary and teaching replacement behaviors to serve the same function of the problem behavior.

Consequences that often work to reduce inappropriate behaviors include ignoring the behavior, verbally and/or physically redirecting the student, reprimanding verbally, taking points or tokens away when using a token economy system and removing the student from the group temporarily. Time-out rooms and physical restraints should only be used in cases of emergency when the student is in danger of getting hurt. Use these with extreme caution and only with permission to do so. The student with Batten disease is often agitated for no apparent reason. If allowed to remain agitated the student sometimes is unable to stop. Therefore, when a function for the behavior and an antecedent to the behavior is unidentified try the following consequences: 1) talk calmly to the student, 2) play music, 3) turn down the lights, 4) hold the student tight, and/or 5) provide time alone in a safe place.

As you can see, behavior management for the student with Batten disease is no easy matter. Rather, behavior management requires a thoughtful, sensitive teacher who analyzes the situations to determine the probable causes before he or she reacts. A functional assessment allows the conscientious teacher to make data-based decisions, a practice recommended by Condon and Tobin (2001).

Ultimately, remember that you are dealing with a special individual who might simply be acting out due to frustration and anger from the disease process. Proceed with caution and with respect. Perhaps the child is upset over conditions beyond his or her control, as when he or she becomes incontinent, evoking feelings of shame and embarrassment (von der Dunk and de Jong, 1992). Perhaps verbal outbursts can be minimized by simply readdressing the content of the lesson rather than the behavior that may be caused by the chaos the child is experiencing in his or her brain (Juvenile Batten's Disease, 1999). As can be seen, behavior management for the student with Batten disease requires great thoughtfulness on the part of the teacher.

Johnston (2001) offers several suggestions when dealing with problem behaviors and the child with Batten. First, "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" (p. 6). Next, remember the child becomes emotionally volatile. Try to reduce occasions when high levels of excitement are likely to occur or avoid too much build-up to upcoming activities. Lastly, provide opportunities for regular relaxation.

Build into the child's routine or schedule times for music, swimming, gentle exercise, massage, aromatherapy, and pets.

Managing problem behaviors can be challenging for the best of teachers. However, you, as the teacher, now have many tools to enable your student to more successfully handle the rough times that evoke misbehavior. Stay calm. Stay flexible. Stay kind.

Individualized Education Plan (IEP)

The Individuals with Disabilities Education Act guarantees students a free and appropriate public education (FAPE). FAPE, for students with special needs, is a program designed to provide educational benefit. Each student's plan is documented in an Individualized Education Program (IEP). Present levels of academic achievement and functional performance (PLAAFP) are expressed in objective, measurable terms to the extent possible. Short-term objectives or benchmarks are stated as a series of achievable components of annual goals that build upon each other over the course of the school year. A statement of related services and supplementary aids and services required must also be included in the IEP.

IDEA mandates a statement of the student's participation in the general curriculum. All students must be assessed, and the IEP documents modifications necessary for the student to participate in statewide and district-wide assessments or if an alternate assessment is more appropriate. The IEP also documents how progress towards goals will be measured and how parents will be informed of this progress. All students age 16 and older must have a transition plan as part of the IEP.

Present levels of academic achievement and functional performance

Koehler and Loftin state that the success of an educational program for students with Batten disease depends on the staff's ability to observe current levels of skills and develop realistic expectations and activities (p. 327). This observation and development process is ongoing as a continuing series of losses, rebounds, and plateaus takes place with all forms of Batten disease (Johnston, p. 14). Thus, begin the IEP process with accurate, data based statements of present levels of academic achievement and functional performance (PLAAFP).

PLAAFPs are often written with both strengths and limitations of the student as the basis. However, focus on strengths and always look for the positive angle when considering PLAAFPs for the student with Batten disease. In other words, focus on the next positive activity rather than on the last negative one (Juvenile Batten's Disease, p. 78). Students with Batten disease are aware of their limitations and remember the time when this was not the case. Von der Dunk and de Jong report, "The youngsters will be able to bear limitations more easily if carers succeed in working with their strong points and interests. The joy of achievement must take priority over the production aspect" (p. 48).

Von der Dunk and de Jong also caution that students may purposely perform below their potential to hide their awareness of limitation (p. 49). If this is the case, the teacher must respond with understanding. Again, focus on strengths and interests, enabling the student to renew his or her motivation to achieve.

Annual goals and short-term objectives or benchmarks

As stated by Martella, “The heart of the IEP includes *measurable* annual goals and short-term objectives or benchmarks that describe each student’s expected learning outcomes. You use annual goals to estimate what *outcomes* you can expect in an academic year based on the student’s present level of performance” (p. 52). In other words, you estimate how much progress the student will make over the course of the year. This estimation is often difficult when dealing with a degenerative condition. For instance, loss of function is inevitable, but the speed of this degeneration is highly variable (Koehler and Loftin, p. 319). Therefore, target goal areas that will have a long-term, functional effect on the child’s life (Koehler and Loftin, p. 325). Suggested goal areas or domains include communication, motor skills, social skills, and daily living or functional living skills. Academic skills remain important for the student to feel challenged and to experience achievement. Adapt the environment and the materials as necessary to challenge the student, programming always for success. According to Koehler and Loftin, “The failure to do so may cause the child to feel no sense of mastery and to become even more depressed at the changes that are occurring” (p. 321).

Annual goals are then divided into short-term objectives or benchmarks, which are intermediate, measurable steps that lead to mastery of the annual goal. Short-term objectives or benchmarks can be sequential skills or can be component skills within a curriculum domain, not necessarily accomplished in sequence (Martella, p. 56-57). A typical example of sequential skills may be to add the assistance level from a most-to-least hierarchy. Consider, though, the reverse of this assistance hierarchy during periods where the disorder develops and the student’s capacity to compensate declines (von der Dunk and de Jong, p. 48). Then, implement a least-to-most hierarchy, remembering that the important aspect of the goal is for the student to achieve TODAY. As an example, last month Dusti could transfer from the mat on the floor to her wheelchair with only a verbal prompt. Today, though, Dusti requires physical prompts along with the verbal prompts to transfer from the mat to the wheelchair. Dusti is as successful today as she was last month on the mastery of transferring from floor to chair. The teacher adjusted her own goals and expectations of Dusti to become more compatible with her capabilities at this time. Stated by Johnston, “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” (p. 8). An example of component skills may be rote counting, one-to-one correspondence, and money identification as component skills in the math domain.

All goals, short-term objectives, and benchmarks should be written as clearly defined and observable behaviors. Martella stresses to write in measurable terms, describing what the student actually will do (p. 55). A well-written goal includes the conditions (i.e., Assistance, setting, etc.), the student’s name, the observable behavior, and the performance criteria. Write them with enough detail so that teachers and parents can implement the goal consistently and know when mastery level is attained or when the goal must be revised. The performance criteria include the percentage correct or rate of correct responding the student demonstrates and how consistently the student performs this rate to consider mastery (Martella, p. 55). An example of a correctly written goal is: When given five calendar objects, Clay will correctly identify the corresponding activity for each item four out of five times for three consecutive performances. Data is then taken regularly to enable the teacher to know when to either add another object to the field of five or to revise the goal.

IEP participants

The following individuals must be present at all IEP meetings:

- Parents
- The child's special education teacher
At least one regular education teacher of the student (if the child may be participating in the regular education environment)
- Local education agency representative (LEA)
- Individual(s) who can interpret the instructional implications of evaluation results (i.e., psychologist, therapist, nurse, etc.)
- Representatives of other public agencies if the child is sixteen years old or older
- The child (if appropriate)

In addition, schools are required to permit the attendance of other individuals invited by the parents and are also permitted to invite other individuals.

Special education law designates IEP participants to emphasize the need of a team approach to special education. This team effort or holistic approach is necessary to provide multi-disciplinary decision making for the physical, mental, emotional, and social needs of the individual child. As a team, consider the child as a member of a family, a peer group, and of society. Also, consider the needs of the family (Juvenile Batten's Disease, p. 107). The IEP team meets at least annually, but the parents may request an IEP at any time (de Bettencourt, p. 20).

Communication between IEP participants is the key to a successful partnership when designing and implementing an IEP. Each member must understand the implications of Batten disease and be informed of any changes that occur throughout the IEP period. Johnston states, "Every parent of a child with Batten disease needs a network of supportive people who have an understanding of the condition" (p. 9). The IEP team provides this network to parents and all other members of the team. Parents should feel welcome to visit the school at any time and the staff should be open to discussing the needs of the parents as well as those of the child (Juvenile Batten's Disease). The journey of living with, caring for, and educating a child with Batten disease is a stressful and emotional journey, but one of the most rewarding journeys you will experience if you maintain an open and caring team approach. Special education law requires that you inform the parents about their child's progress at least as often as parents of students without disabilities. This time schedule is often interpreted as a quarterly reporting schedule. However, this time schedule of reporting progress may not be adequate for the student with Batten disease. Rather, daily or weekly reporting may be necessary. Actually, Johnston recommends a notebook that travels between home and the school to record any changes in medications, moods, skills, motivation, etc. This system of communication is highly recommended. Then, any individual interacting with the child can record progress or problems in the notebook to keep all IEP team members informed on a regular basis.

Realistic expectations

Referring to students with Batten disease, Koehler and Loftin state, “Maintaining a realistic level of expectations for the child may be one of the most difficult tasks that a teacher faces” (p. 323). As you develop goals, short-term objectives, and/or benchmarks the following points will aid you in maintaining a realistic level of expectations for your student with Batten disease:

- Remain positive
- Teach to the child’s strengths and interests
- Teach functional skills
- Adapt the curriculum and materials so as the child can continue to achieve
- Provide a high level of stimulation, both mental and physical
- Remember that the child can still recall his or her normal existence
- Provide age appropriate activities
- Respect the child’s rights and needs in all stages of the disease
- Stress quality of life now
- Have fun!

Many teachers appear to have difficulty in writing goals when the student is actually regressing in one or more areas. Be creative and flexible. Observe, observe, observe, and adapt, adapt, adapt! Von der Dunk and de Jong give us an accurate perspective: “Enjoying school is more important than completing certain syllabuses. We are not talking about material designed to prepare a pupil for a job or a place in society, but about acquiring fruitful experiences now. Fun and finding pleasure in being occupied are an important counterbalance to stress” (p. 47).

As an example of how to scale back an IEP to maintain realistic expectations, let’s follow Emily through her educational experience. Emily began kindergarten in a regular education classroom in her neighborhood school. Then, Emily attended a special school for the blind and visually impaired for seven years. At age twelve, Emily transferred to a center-based school for students with multiple disabilities. Her first IEP at this school included the following goals:

- Passive range of motion
- Transferring from a tall kneel to a half kneel
- Using lateral tongue movement while eating
- Pointing to pictures on an augmentative system
- Signing 30 words
- Answering comprehension questions
- Sorting into four to six groups
- Completing a worksheet
- Following directions involving spatial concepts
- Signing ten reading words
- Stringing beads from one to ten
- Counting up to ten blocks
- Saying the next number from one to twenty

During Emily’s second year at this school she was maintaining her gross motor physical abilities quite well. However, her communication, fine motor, and academic skills were declining.

Therefore, her teacher continued the physical goals of range of motion and transferring from a tall kneel to a half kneel, but adapted most of her other goals to be more functional and important to Emily. This IEP included the following goals:

- Passive range of motion
- Transferring from a tall kneel to a half kneel
- Pointing to pictures on an augmentative system
- Answering questions using one to three signs or words
- Answering yes/no comprehension questions
- Identifying personal information from two verbal distracters
- Signing ten menu words
- Counting dollars or coins from one to ten
- Saying the next dollar when told a price
- Wiping tables
- Sorting laundry
- Sorting silverware
- Participating in weekly community-based activities

Emily's third year at this school was in the same class as the previous two years. Her skills remained fairly consistent, but her agitation level began to increase. The focus of this school year was to maintain her skills as much as possible and to continue to teach functional skills. Emily's IEP this year included the following goals:

- Passive range of motion
- Transferring out of her wheelchair
- Walking to the lunchroom
- Hitting a switch to access an augmentative device
- Answering questions
- Complying with "Put hands in lap"
- Ordering a food item
- Selecting two items in the grocery store
- Asking for assistance in a convenience store
- Showing I.D. tag upon request
- Washing face
- Washing hands
- Brushing teeth

The disease process took its toll during this year that resulted in Emily moving to another classroom where it was quieter yet still stimulating. Emily appeared much more content in this environment and was able to once again progress on her IEP goals. However, her goals were scaled back and were adapted to enable Emily to achieve success. This year's IEP goals were as follows:

- Passive range of motion
- Ambulating in the swimming pool
- Standing in the prone stander
- Completing a fine-motor task
- Washing hands

- Answering questions with an augmentative device
- Using a tactile symbol system

Emily continued to appear content during her fifth year. However, her teacher had to scale her goals back even further and adapt by increasing the assistance level that Emily required to complete her tasks. Emily remained stimulated and happy because she was involved in enjoyable activities, both actively and passively, and she felt successful. Her goals for this year were as follows:

- Passive range of motion
- Opening her calendar boxed
- Identifying calendar box objects
- Raising hand to signal 'finished'

Emily's fifth year at this particular center-based school turned out to be her last year. She remained happy and content due to the love and respect that the staff had for her. The focus was always on a positive approach. When a skill was lost or too difficult it was adapted or replaced by a more appropriate skill that could be accomplished.

One final caution concerning the above example of IEPs: Remember that every student is an individual. The individual progress and the disease process will vary. The examples are given as a springboard to your own thinking when developing an IEP for your special student.

Enjoy your journey through the years with your student with Batten disease. The journey will be like no other providing that you remember what is happening throughout the disease process. Von der Dunk and de Jong add, "Children who suffer from dementia can still recall their normal existence. These memories tend to disintegrate and consist of vague flashes, but find no comparison in mentally deficient children with similar intelligence levels" (p. 17). Your student with Batten disease is unique and will respond to the respect and stimulation that you can provide.

Your student with Batten disease will provide you with challenges. Writing IEPs will require your best knowledge, creativity, and flexibility. Implementing those IEPs will require more of the same. However, as Koehler and Loftin state, "Children with such unique medical conditions present some of the most challenging yet rewarding aspects of work in special education" (p. 327).

Parental Participation

To protect parents' rights in the IEP process, the school must provide notice, schedule for mutual convenience, and facilitate understanding of the process.

As directed by the Office of Special Education Programs, the content of the notice must include the purpose, time and location of the meeting, and who will be in attendance (sufficient to indicate by position only).

According to Cordrey v. Euckert, 17 EHRLR 104 (6th Cir. 1990), parents' unwillingness to participate will not invalidate the IEP if attempts to arrange a mutually convenient meeting and convince the parents to attend are documented.

Parents must also be involved in re-evaluation assessments and decisions.

Parental participation in placement decisions is mandated under Special Education law, regardless of whether those decisions are made by the IEP team or some other group.

Encourage parental participation through daily notebook communication. It is important for caregivers in all settings to know how the child ate, seizure activity, special events that occurred, etc. Compassionate and sincere responsiveness to all parental participation is critical to the family and to the student. They need the school's support. In return, the family will indeed appreciate the extra effort taken for their child.

Placement in Special Education

The team making placement decisions must include, of course, the parents. The team making placement decisions must:

- Draw upon information from a variety of sources
- Ensure that information is documented and carefully considered
- Ensure that the placement decision is made in conformity with the least restrictive environment (LRE) rules

Special Education law creates a preference for neighborhood schools. However, the court in Murray v. Montrose County School District, RE-1J, 22 IDELR 558 (10th Cir. 1995) found that Special Education law does not create a presumption in favor of neighborhood schools and therefore does not necessarily create a right to placement there. School districts are to ensure that a continuum of alternative placements is available.

The continuum of alternative placements begins with full-time placement in the regular education classroom as the least restrictive environment. Following is a complete list of educational placement options with each level signifying a more restrictive environment:

- Regular classroom
- Regular classroom with special education consultant
- Regular classroom with itinerant teachers
- Regular classroom with use of resource room for part of the time
- Part-time special class
- Full-time special class
- Special day school
- Residential school
- Home or hospital (Rothstein, p. 135).

McLeskey, Henry, and Hodges report, “One of the most significant changes that has occurred in public education in the United States over the past 15 years has been the movement toward inclusion – that is, educating students with disabilities for increasingly more of the school day in general education classrooms” (p. 4). However, the decision to place a child in an inclusive setting or a more restrictive special education program is seldom a clear-cut decision. There are several important factors to weigh when considering educational placement decisions, including the quality of the available programs, the availability of specialized services, and access to family-centered approaches, as reported by Hanson, et al. (p. 66). Importantly, Rothstein reminds us that the nature of these placement decisions is fluid. In other words, the placement must be reviewed at least annually to verify that the educational alternative best serves the individual student at this time (p. 135). For the student with Batten disease, changes will occur, requiring constant assessment of the appropriateness of the educational placement. Therefore, consider appropriateness as well as least restrictive especially for these special students.

Inclusion

A study by Hanson, et al. reports parents favoring inclusion to help their children reach their potential for socialization, to make friends, and to provide role models for behavior, speech, and social skills (p. 73). Another important determining factor is for the child to be happy. "Parents in this study generally advocated for inclusive placements until they or their children encountered obstacles at school" (p. 77). The parents' primary concerns when deciding to maintain their child's placement in inclusive settings or to move to a more restrictive setting include:

- Class size
- Availability of specialized therapies and services
- Children's acceptance by others
- Teachers' judgments or attitudes about the child's disability
- Parents' judgments of the appropriateness of teachers' training and experience in addressing the children's needs (p. 78).

Many students with Juvenile Batten Disease begin their educational experience in inclusive settings and remain there for quite some time. As visual impairments worsen and other disease related impairments become apparent the above concerns will eventually need to be addressed.

However, the following factors, presented by Cook, Tankersley, Cook, and Landrum, may increase the success of the inclusive educational setting:

- An extra instructional assistant
- The teacher's experience teaching in inclusive classrooms
- The teacher's training in special education and inclusion
- Time for the teacher to collaborate with special education personnel outside of the class
- Presence of special education personnel in the inclusive classroom
- Lower class sizes (p. 118).

The move to a more restrictive educational placement

In the initial phase of Juvenile Batten Disease when there is still little evidence of mental degeneration, the children become aware of failing eyesight and difficulty keeping up in school. Regular education to special education at this time may make the children feel like outcasts (von der Dunk and de Jong, p. 19). However, the material and pace at the school will become too difficult. The children become painfully aware of their shortcomings. Hofman reports a decrease in the ability to change one's attention quickly from one subject to another, to learn new skills, and to put forth effort. The tempo of things happening around the student becomes too fast, resulting in a loss of grip on reality. Irritability and changes of mood accompany this process. The realizations by the student that body functions are deteriorating and that epileptic symptoms and involuntary movements are occurring often result in anxiety and confusion. This anxiety is sometimes accompanied by threateningly tinted delusions and hallucinations (p. 66).

Whether periods of excitement, anxiety, or delusions are a result of environmental factors, problems in coping, organic causes, or a combination of these factors, a more peaceful environment lacking in stimuli may improve the behavior (Hofman, p. 68). **At this time, the best educational placement may very well be to a more restrictive setting.** Decisions become based on a process of letting go. Johnston states, “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” (p. 10). Von der Dunk and de Jong add, “After a phase of confusion, resistance and denial, the child usually experiences a kind of acceptance, acquiescence or resignation. Acceptance of the condition can give a feeling of relief, of peace: ‘I don’t have to do that anymore’” (p. 49).

Amber, a teen with Batten disease, attended school in a special classroom where the noise level, activity level, and expectations were high. Amber loved school but became increasingly agitated throughout the school day. She cried a lot, screamed out, and refused to try tasks that were within her ability level. The IEP team, including her mother, made the decision to move Amber to a class for students with more significant disabilities. The environment was much quieter, the pace much slower, and the expectations consistent with the abilities of the individual children. Amber calmed down within days in this placement. Her seizures decreased, and she smiled a lot more. She was happy! Amber remained in this educational placement for the remainder of her educational experience.

Amber’s educational placement was at a center-based school. Center-based schools provide a centralization of services, increasing the proficiency of personnel and maximizing the availability of staff. This may be a factor in some districts, as cost becomes an issue when serving students with low-incidence, high-cost disabilities. Regardless of the justification of and need for center-based schools, the IEP must now include an explanation of the extent, if any, to which the child will not participate with non-disabled children in the regular class and in the general education curriculum including extra-curricular and nonacademic activities.

A need for continuity

Children with Batten disease benefit highly from as much continuity as possible. Repeated change is detrimental to their peace of mind and quality of life. Therefore, concerning placement decisions, von der Dunk and de Jong state, “If it looks to be a brief, temporary move, it will demand a lot of energy, but will also mean even greater disappointment and dejection when the child has to be transferred again” (p. 47). Make placement decisions carefully, considering all the possible implications to the individual student. Along with continuity, recommend an educational placement that provides a loving environment, appropriate stimulation, and care as individuals (Juvenile Batten’s Disease, p. 69).

Students with degenerative conditions such as Batten disease present challenges to the educational system that is typically based upon progression rather than regression. However, with the options available to districts, IEP teams are able to make individualized and *appropriate* placement decisions to benefit the student with Batten disease and his or her family. Each student is unique and has different needs. **The bottom line, though, is for the student with Batten disease to be happy in his or her educational placement.**

Related Services

Related services are developmental, corrective, or supportive services required to assist the child to benefit from special education. Related services are defined as:

Transportation, and such developmental, corrective, and other supportive services (including speech-language pathology and audiology services, psychological services, physical and occupational therapy, recreation, including therapeutic recreation, social work services, counseling services, including rehabilitation counseling, orientation and mobility services, and medical services, except that such medical services shall be for diagnostic and evaluation purposes only) as may be required to assist a child with a disability to benefit from special education, and includes the early identification and assessment of disabling conditions in children (Special Education Law).

Parents, teachers, and therapists collaborate as a team to assess the student and his or her related service needs, write goals, and implement intervention (Barnes and Whinnery, 2002). It is important to note that the implementation of goals, along with the necessary related services, be embedded within the student's natural daily activities (Barnes and Whinnery, 2002).

Transportation

Transportation to and from school is a related service. The duty to provide transportation as a related service begins at the curb or at the door depending on your particular district. The need for health-related services during transportation does not relieve the district of the responsibility of providing transportation as a related service.

Speech-language pathology

A speech-language pathologist typically becomes involved in the education of students with Batten disease when difficulties with speech patterns become apparent. However, speech-language services should also continue as speech ceases. For example, the speech-language pathologist coordinates and implements calendar systems that will enable the student to communicate when he or she is no longer able to verbalize. One vital component of speech-language services for students with Batten disease is the introduction and utilization of objects as representations of activities early in the disease process and the consistent continuation of this intervention throughout the remaining educational process.

A study by Lamminranta, Aberg, Autti, Moern, Laine, Kaukoranta, and Santavuori (2001) revealed that as cognitive ability declined due to the disease process, recognition of similarities and information remained the best abilities during the study period. Reported by Lamminranta, et al., "Comprehension of speech remained unchanged. Auditory perception and phonemic discrimination, the ability to discriminate between sentence structures and phonemes, remained quite stable" (p. 13).

An assistive technology device may be a related service, special education, or a supplementary aid and service. Special Education law requires the IEP team to consider whether the child requires assistive technology devices and services.

Audiology services

Audiology services as related services include hearing assessments at least every three years and the provision and implementation of services to aid the student that has hearing deficiencies.

Psychological services

The school psychologist assists in evaluations, behavior assessment and intervention planning, and aids the student when personal difficulties arise that negatively affect the educational and emotional processes. Anger management may be an area where the psychologist can provide assistance. Koehler and Loftin (1994) report that anger often exists towards the steadily increasing dependence on others experienced by the student with Batten disease.

Physical and occupational therapy

The physical therapist focuses primarily on gross motor functioning, such as walking. **Mobility is important as long, and only as long, as the activity remains pleasant.** As walking becomes difficult and stressful, perhaps the physical therapist could move therapy sessions to the swimming pool where the buoyancy of the water allows the student to continue to be successful with a skill that is declining. Also, if the student enjoys physical education (PE) class but hates private therapy sessions, perhaps the therapist could provide services during PE class and embed therapy into the activities enjoyed by the student.

The occupational therapist focuses primarily on fine motor functioning. As fine motor functioning declines, the occupational therapist adapts the activities so as to allow for continued success. For example, the student that loves to string small beads to make necklaces eventually finds this activity frustrating and begins to lose interest in this once mastered skill. The wise occupational therapist will adapt this activity by providing large beads with large holes and stiffer string or pipe cleaners to string them on to allow the student to once again find success in a skill once enjoyed.

Range of motion is another aspect of physical and occupational therapy that the therapists will try to maximize and maintain. This therapy should be done slowly and comfortably. The key is to not increase the stress level of the student. If the student becomes anxious, perhaps the therapist could try therapy sessions in a dimly lit room with pleasant music playing and other sensory stimulation available as enjoyed by the student. **Passive range of motion should cease if the activity causes stress and agitation to the student.**

Recreation

Recreation is important for each and every one of us. Recreation as a related service may be in the form of teaching and providing leisure skills that the student enjoys. Adapted physical education is another form of recreation as a related service.

Social work and counseling services

Social work and counseling services work hand in hand with psychological services. These services can aid the student with Batten disease with social interaction and social cooperation as peer interaction typically changes and declines as the disease process continues. Intervention may include teaching simple board games, facilitating group art projects, providing assembly line cooking, and engaging in role-plays of various situations, both fun and problematic (Sadler, 2001).

Koehler and Loftin (1994) report, "It is critical for the counselor to meet regularly with the staff to reinforce the need to maximize opportunities for the child's independence in his or her day-to-day life for as long as possible" (p. 326). This communication should also extend to the family. In addition, the social worker or counselor can also be a valuable resource to the family when researching available services beyond the school setting (i.e., Respite, Medicaid, Social Security Income, etc.).

Orientation and mobility services

Orientation and mobility is the process of knowing where you are in relationship to others and objects in the environment and the ability to move from one place to another in this environment. Orientation and mobility (O&M) is often thought of as one because orientation provides the motivation to move. In a study by Lamminranta, et al., (2001), the authors found that orientation to time declined more than orientation to people and places.

The O&M specialist will work on traveling skills as long as ambulating is appropriate. However, the O&M specialist can still provide services after the student has lost this ability and has become wheelchair bound. For example, the O&M specialist can help the student maintain his or her orientation skills allowing for the student to know where he or she is in the classroom, school, and surrounding environments. One suggestion is to attach an identifying object to the door of various locations in the school. The student can then determine what area of the school or what classroom he or she is entering. This will promote a feeling of independence during a time when independence is becoming limited.

Vision services

Students with Batten disease may likely have a vision specialist as well as an O&M specialist. The vision specialist assesses the student's vision and consults with the classroom staff concerning issues to aid the student's ability to use what vision is available. Ross (2002) provides a number of suggestions to aid the student who has visual impairment. Following are suggestions that may benefit the student with Batten disease:

- Use names when calling on students in class
- Use reference points such as north, south, etc. or the numbers on a clock
- Refer to objects using several attributes such as color, shape, weight, texture, size, and location
- Check for understanding
- Teach the layout of the classroom and notify student if it changes
- Use front row seating in the classroom and at assemblies
- Ensure a view of the board that is free from glare – allow tinted glasses, hats, or visors
- Teach while standing away from the windows
- Monitor the lighting in the classroom
- Pair students up to work together
- Use white dry-marker boards
- Xerox teacher's or other student's class notes
- Allow student to preview visuals and manipulatives to be presented during instruction
- Talk as you write on the board
- Xerox copies of overheads for the student to have at his or her desk
- Be descriptive and precise during instruction
- Xerox spelling lists, homework assignments, etc.
- Use high contrast and enlarged size as appropriate

Medical services

The court in Irving Independent School District v. Tatro (1984) established a four-part test for distinguishing whether a particular health related service is covered under Special Education law. Accordingly, the limitations include:

- The student must be IDEA-eligible
- The service must be necessary for the student to benefit from special education
- The student must need to be provided with the service during school hours
- The service can be performed by a non-physician

Important! Simply stated, health-related services that do not require a physician or hospital to administer are medical services that may be considered related services that the school district must provide. School health care, as a related service, enables many medically fragile children to attend school. Concerning a paralyzed and ventilator-dependent adolescent in Cedar Rapids Community School District v. Garret F. (1999), the court stated that the lack of stimulation the student now receives from teachers and peers would have a profound effect on the rest of his life.

The administration of medication may be a related service. Essentially, any health-related service that a nurse can perform or delegate to a properly trained layperson is not an excluded medical service. This is true regardless of the cost of the service or the burden it creates on a school district's resources.

Any related service deemed appropriate for the student with Batten disease will only be as successful as it is pleasant. Make education as enjoyable for the student as possible. The results will be rewarding to the student, to the family, and to the teacher.

Procedural Safeguards

Procedural safeguards include the following:

- Parents can examine all records, participate in identification, evaluation, placement, and decisions to provide a free, appropriate education (FAPE), and obtain an independent educational evaluation
- Provision of surrogate parent if necessary
- Written prior notice whenever the school: 1) proposes to initiate or change, or 2) refuses to initiate or change the identification, evaluation, placement, or the provision of FAPE
- Notice provided in native language of the parents
- Opportunity for mediation
- Opportunity to present complaints
- The procedures to provide notice when complaints are filed
- Assistance in filing a complaint

Special Education law defines a 'parent' as a parent, guardian, person acting as a parent of the child, or an appointed surrogate parent. The definition does not include the State if the child is a ward of the State.

The contents of the prior written notice must include:

- Description of action proposed or refused
- Explanation of why
- Description of other options considered and reasons why rejected as basis for proposed action
- Description of any other relevant factors
- Statement of parents' rights and how to obtain a copy of the description of the procedural safeguards
- Sources for parents to obtain assistance in understanding the provisions

In addition to prior written notice for identification, reevaluation, and placement, the school must also obtain parental informed consent prior to conducting a reevaluation. To confirm that consent is informed, the parent must sign a written consent form that describes the action to be carried out. Consent is voluntary and may be revoked at any time.

Notes for the School Nurse

School nurses are playing a bigger role in the school setting each year. Also, the school nurse has become a member of the medical staff for each individual student. Nursing has become more technical and scientific as well as being the nurturing phase of the medical care. The school nurse assesses and intervenes with students to treat, prevent, and educate, and to help people cope with various health care situations. The nurse must deal with the physical, emotional, intellectual, and spiritual aspects. The nurse, as well as the teachers, are key players to assess the student's physical, social, and emotional aspects, to administer therapy and medications, and to teach the student, family, and caregivers how to cope with the therapy. To ensure the most effective therapy and evaluate that effectiveness requires a broad base of knowledge in the basic sciences (anatomy, physiology, nutrition, chemistry, pharmacology), the social sciences (sociology, psychology), and education (learning approaches, evaluation).

The practice of nursing includes the nursing process with key elements of assessment (gathering information), diagnosis (defining that information to arrive at some conclusions), intervention (administration, education, and comfort measures), and evaluation (determining the effects of the interventions that were performed). The nursing process also helps parents keep accurate and up-to-date records as part of the educational team.

Health care plans

Some of the health concerns that may require health care plans at school are the following:

- **Seizures** – Have a plan in place detailing the family's wishes about CPR, status epilepticus, and how long to wait until 911 is called (i.e., 3 minutes, 5 minutes). Update protocols with increasing occurrence of seizures. If seizures are not currently manifesting themselves in a student, they will in the future, and the frequency will also increase. The seizures may be of different types: Generalized, tonic-clonic (grand mal), tonic, clonic, absence (petit mal), atonic (drop seizures), partial seizures (which can be further broken down to simple partial, call focal seizures, and complex partial, called psychomotor seizures), myoclonic, temporal lobe (Jacksonian seizures, which begin with partial seizures and extend to generalized seizures). Have phone numbers of the neurologist, pediatrician, and parents easily accessible. Patterns of onset, factors, frequency, and triggers will vary from child to child. The description of a seizure, including timing of a seizure, is also very important as a medical tool for diagnosing and for administering medication dosages or making adjustments in medications.
- **Progressive vision loss** – The child will need increased orientation training with increasing vision loss and assistance with all activities. Issues such as public schools versus blind schools as well as learning Braille versus being read to need to be addressed.

- **Impaired physical mobility** – Clumsiness, staggering, wheelchair use, complete dependence, skin care, respiratory function, and bowel and urine difficulties will likely be experienced. A ‘bent-knee to toe-in’ posture is typical for children with Batten disease who are still ambulatory. Ambulation, movement exercises, and support to prevent falls and injuries need to be considered with each child.
- **Impaired speech** – Increased difficulty with articulation, repeating sounds, syllables and words over and over, slowness in processing information, and delay in answering indicate the need for accommodation.
- **Nutrition** – The child needs to be evaluated for swallowing safety, the inability to eat typical foods, the need for alternative feedings or Gastrointestinal tube placement to avoid aspiration, and the need for suctioning. Nutritious meals and adequate fluids are essential. Gastric reflux and drooling can become issues for some of the children.
- **Loss of body function** – Disturbances in self-concept may be evident with incontinence of urine and stool. Also, constipation is a problem that needs to be addressed for many children with Batten disease.
- **Dementia** – Loss of control, memory loss, behavioral outbursts, and hallucinations are often due to dementia. Encourage parents to seek medical counseling for medications that may help decrease symptoms.
- **Medications** – A list of the child’s medications (dosages and times to be given), recent blood levels, weight increases or decreases, and break-through or loss of control of seizures is helpful in the ongoing care of the child.
- **Knowledge of the disease** – The family and caregivers need all of the information they are ready for from as many sources as possible. One tendency of children with Batten disease is the high motivation level in the face of regressing skills that will definitely contribute to frustration for them. Thus, your support and understanding will be most beneficial.
- **General health** – Children with Batten disease are still susceptible to cold, flu, measles, chicken pox, etc. In addition, they may go through puberty early, with girls starting their menstrual periods early.

The above is a brief outline of a few medical aspects of Batten disease. For more information contact Nancy Carney, RN and Medical Liaison/Educator, at 1-877-642-5512 or e-mail nancycarney@bdsra.org.

Make Time Count!

You, as the teacher of a student with Batten disease, can enhance the quality of life for your student now and in the future. What you do today can make a difference for the rest of your student's life. With knowledge of the disease and the importance of the instruction you provide, you can make time with your unique and special student count. The journey is just beginning for you. Make it a rewarding one for your student, for his or her family, and for yourself. The lessons you learn from your student with Batten disease will enable you to become a better teacher to all students that you encounter in your teaching career.

Good luck and God bless. Make it count!

The BDSRA values your experience and expertise. If you have any ideas, suggestions, or interventions that work, please let us know! Also, for further information contact:

Batten Disease Support and Research Association
166 Humphries Dr., Reynoldsburg, OH 43068
800-448-4570

How to Contact Us

Contact persons:

Wendy Bills, Chair, BDSRA Education Committee	Email: wbills@murrayschools.org
Margie Frazier, PhD, Exec. Director, BDSRA	Email: mfrazier@bdsra.org
Lisa Weston, MPA, Program Director, BDSRA	Email: lisaweston@bdsra.org

Educational Consultants (available by phone, e-mail, or in person for inservice at your site for expenses only – no speaker or consultant fee):

Lindsey Adams (Utah)		Email: ladams@murrayschools.org
Wendy Bills (Utah)	866-287-7232 (toll-free)	Email: wbills@murrayschools.org
Emily Calvert (Texas)	281-989-7412 (cell)	Email: kevin.calvert@sbcglobal.net
Jane Farber (Maryland)		Email: Jane.Farber@pgcps.org
Colleen Heath (Ohio)		Email: cheath@ossb.oh.gov
Toni Hollingsworth (Mississippi)		Email: hollingsworth.toni@gmail.com
Chris VanderStel (Michigan)		Email: cvanderstel@fhps.net

BDSRA wishes to express sincere appreciation to the following contributors:

Wendy Bills, Ph.D., Special Education Director, Murray School District, Murray, UT
Lance W. Johnston, Parent and Former Executive Director of BDSRA
Nancy Carney, R.N.
Amy Kirk
Bob Wilhelm, M.S., Parent
Madelyn Gardner, Physical Therapist, Ohio State School for the Blind
Leslie Graham, Ph.D., Bradley University, Peoria, IL
Christy Hubbard, M.Ed., Developmental Disability Teacher, Ohio State School for the Blind
Judy O'Keefe, Ph.D., Speech/Hearing Pathologist, Ohio State School for the Blind
Debbie McClendon, B.S., Special Education Teacher, Harrison Co. School Dist., MS
Lindsey Adams, M.S., Special Education Teacher, Murray School District, UT
Emily Calvert, Special Education Teacher, Cy-Fair School District, Houston, TX

*A special thanks to Lori Wenerstrom for her artwork (Figures 1-4.)
Email: pencilpointsrt@gmail.com*

A very special thanks to my husband, Clay Bills, for all his patience and support. Without him, my years of service with BDSRA could not have happened. I also want to thank him for his artwork (Figures 5 and 6). ~ Wendy Bills

REFERENCES

- Barnes, S. B. & Whinnery, K. W. (2002). Effects of functional mobility skills training for young students with physical disabilities. *Exceptional Children*, 68, 313-324.
- Blaha, R. & Moss, K. (1997). *Let Me Check My Calendar*. <http://www.tsbvi.edu/Outreach>.
- Brown, D. *Resonance Boards*. California Deaf-Blind Services.
- Condon, K. S., & Tobin, T. J. (2001). Using electronic and other new ways to help students improve their behavior: Functional behavioral assessment at work. *Teaching Exceptional Children*, 34(1), 44-51.
- Daniels, V. I. (1998). How to manage disruptive behavior in inclusive classrooms. *Teaching Exceptional Children*, 30(4), 26-31.
- De Bettencourt, L. U. (2002). Understanding the differences between IDEA and section 504. *Teaching Exceptional Children*, 34(3), 16-23.
- Downing, J. E. & Peckham-Hardin, K. D. (2001). Daily schedules: A helpful learning tool. *Teaching Exceptional Children*, 33, 62-68.
- Dunnett, J. (1997). Nielsen's Little Room: It's use with a young blind and physically disabled girl. *Journal of Visual Impairment and Blindness*, Mar.-Apr., 145-150.
- Hanson, M. J., Horn, E., Sandall, S., Beckman, P., Morgan, M., Marquart, J., Barnwell, D., & Chou, H. (2001). After preschool inclusion: Children's educational pathways over the early school years. *Exception Children*, 68(1), 65-83.
- Hofman, I. L. (1999). *The Batten-Spielmeyer-Vogt disease*. Bartimeushage, Doorn, Netherlands.
- Johnston, L. (2001). *Batten Disease: Insights for parents*. Reynoldsburg, Ohio. Batten Disease Support and Research Association.
- Juvenile Batten's Disease: Care in Partnership*. (1999). See Ability/Heather House.
- Koehler, W., & Loftin, M. (1994). Visually impaired children with progressive, terminal neurodegenerative disorders. *Journal of Visual Impairment and Blindness*, July-Aug., 317-328.
- Lamminrants, S., Aberg, L. E., Autti, T., Moren, R., Laine, T., Kaukorants, J., & Santavuori, P. (2001). Neuropsychological test battery in the follow-up of patients with juvenile neuronal ceroid lipofuscinosis. *Journal of Intellectual Disability Research*, 45(1), 8-17.
- Lewis, S. & Tolla, J. (2003). Creating and using tactile experience books for young children with visual impairments. *Teaching Exceptional Children*, 35, 22-28.
- Maag, J. W. (2001). Rewarded by punishment: Reflections on the disuse of positive reinforcement in schools. *Exceptional Children*, 67(2), 173-186.
- McLeskey, J., Henry, D., & Hodges, D. (1998). Inclusion: Where is it happening? *Teaching Exceptional Children*, 31(1), 4-10.
- Monaco & Mamer (1993). *The experience book*. Brantford, Ontario. The W. Ross Macdonald School.

- O'Neill, R. E., Horner, R. H., Albin, R. W., Sprague, J. R., Storey, K., & Newton, J. S. (1997). *Functional assessment and program development for problem behavior: A practical handbook*. Pacific Grove, CA. Brooks/Cole.
- Ross, J. (2002). Some specifics for the classroom teacher of blind or visually impaired students. *Division on Visual Impairments Quarterly*, 47(3), 35-37.
- Ross, J. (2002). Suggested modifications for classroom teachers of students with visual impairments. *Division on Visual Impairments Quarterly*, 47(2), 49-50.
- Rothstein, L. F. (2000). *Special Education Law* (3rd Ed.). Addison Wesley Longman, Inc. New York.
- Sadler, F. H. (2001). The itinerant teacher hits the road: A map for instruction in young children's social skills. *Teaching Exceptional Children*, 34(1), 60-66.
- Schreck, K. B. (2002). *Educating the child or adolescent with juvenile neuronal ceroid lipofuscinoses or Batten Disease*. Unpublished master's thesis, North Park University, Chicago, Illinois.
- von der Dunk, M., & de Jong, C. G. A. (Ed.). (1992). *Growing Up to Dependence: Children and Young People with Batten-Spielmeyer-Vogt Disease*. (Available from Bartimeus Foundation, PO Box 87, 3940, AB Doorn, Netherlands).