

# The Illuminator

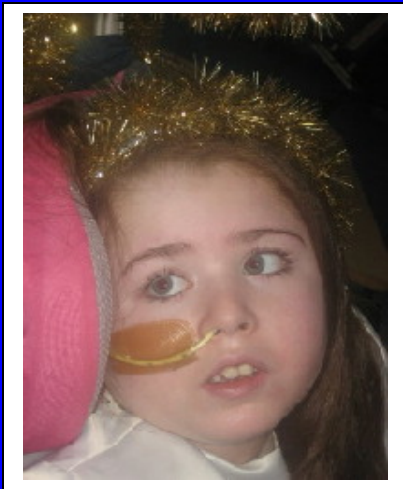
“A light in a world of darkness”

January 2010  
Volume 21, Number 1

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## Special Child



Rhiannon Bates -- LINCL



T-Shirt Designs by JNCL Dad  
Josh Smerdel (story on page 4)

Batten Disease Support &  
Research Association  
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1-800-448-4570  
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## BDSRA CHICAGO 2010



### Field of Hope and Dreams

### BDSRA 2010 Conference Chicago, IL Hosted by Midwest Chapter and National Office

It's officially 2010 and we're already in full swing as we plan for the 22<sup>nd</sup> Annual BDSRA Conference. Be sure to look for the official Conference Pack in the April Illuminator as well as on our website. For now, here are some of the basic details for the 2010 Conference.

Dates: July 29 - August 1, 2010  
 Location: Chicago Marriott Oak Brook Hotel  
 1401 West 22<sup>nd</sup> Street  
 Oak Brook, IL 60523  
 Phone and Reservations: (630) 573-8555  
 Toll-free (800) 228-9290  
 Conference Name: Batten Disease Association Conference  
 Website:  
<http://www.marriott.com/hotels/travel/chiob-chicago-marriott-oak-brook/>  
 Room Rate: \$95.00 per night

Meal prices will be available soon. Please check the BDSRA website for Conference updates.

## BDSRA Approves Juvenile Drug Trial Funding

The Board of Directors of the Batten Disease Support and Research Association is pleased to announce that it has approved a \$400,000 award to the Batten Disease Diagnostic and Clinical Research Center at the University of Rochester for a clinical trial of a drug for Juvenile Batten Disease. The drug, Mycophenolate Mofetil, also known as CellCept, is an immunosuppressant used to prevent rejection of transplanted organs in children. In preliminary studies, mice affected by Batten Disease demonstrated improved motor skills when treated with the drug.

The total cost of the trial is \$1.1 million. The research team, under the direction of Dr. Frederick Marshall and Dr. Jonathan Mink, is working to raise the additional \$700,000 required to begin the study. In the meantime, funds for the Juvenile Trial will continue to be accepted by BDSRA, as more funding could potentially be needed. Any funds designated towards the Juvenile Trial that are not used to fund the trial will be applied to Juvenile research.

*Research updates continue on page 2*

# Research Update

## UI scientists use blood brain barrier as therapy delivery system

The blood brain barrier is generally considered an obstacle to delivering therapies from the bloodstream to the brain. However, University of Iowa researchers have discovered a way to turn the blood vessels surrounding brain cells into a production and delivery system for getting therapeutic molecules directly into brain cells.

Working with animal models of a group of fatal neurological disorders called lysosomal storage diseases, the UI team found that these diseases cause unique and disease-specific alterations to the blood vessels of the blood brain barrier. The scientists used these distinct alterations to target the brain with gene therapy, which reversed the neurological damage caused by the diseases.

The findings, which were published Sept. 13 in Nature Medicine's Advance Online Publication (AOP), could lead to a new non-invasive approach for treating neurological damage caused by lysosomal storage diseases.

"This is the first time an enzyme delivered through the bloodstream has corrected deficiencies in the brain," said lead investigator Beverly Davidson, Ph.D., UI professor of internal medicine, neurology, and molecular physiology and biophysics. "This provides a real opportunity to deliver enzyme therapy without surgically entering the brain to treat lysosomal storage diseases."

"In addition, we have discovered that these neurological diseases affect not just the brain cells that we often focus on, but also the blood vessels throughout the brain. We have taken advantage of that finding to delivery gene therapy, but we also can use this knowledge to better understand how the diseases impact other cell types such as neurons," she added.

Lysosomal storage diseases are individually quite rare, but as a group they affect approximately 1 in 8,000 live births. The diseases are caused by deficiencies in enzymes that break down larger molecules. Without these enzymes, the large molecules accumulate inside cells and cause cell damage and destruction.

Enzyme replacement therapy has been successful in treating one form of lysosomal storage disease called Gaucher disease. However, storage diseases that affect the central nervous system remain untreatable because it has not been possible, to this point, to get the missing enzymes past the blood-brain-barrier and into the brain.

"Our discovery allowed us to test the idea that the brain cells might be able to make use of the reintroduced enzyme to stop or reverse the damage caused by the accumulated materials," said Davidson, who also is the Roy J. Carver Professor in Internal Medicine. "In the treated mice, the affected brain cells go back to looking normal, the brain inflammation goes away and the impaired behaviors that these mice have is corrected."

To develop their gene therapy targeting system, Davidson and colleagues used a technique called phage panning to identify peptides that hone in on the blood vessels surrounding the brain. Surprisingly, they found that peptides that targeted the brain blood vessels in mice with lysosomal storage diseases were distinct from the peptides that targeted brain blood vessels in healthy mice. Moreover, the peptides that targeted blood vessels in different diseases were distinct from each other, suggesting that each disease causes specific alterations to the blood vessels.

The team modified a deactivated virus used for gene therapy so that the virus expressed copies of the unique brain-targeting peptide on its outer coat, and also carried the genetic blueprint for the missing enzyme.

The study showed that the modified virus targeted the blood vessels in the brain and caused the blood vessel cells to produce the enzyme. Most importantly, the researchers found that the enzyme was secreted into the brain tissue in sufficient quantities to correct the disease symptoms and problems.

The team was able to use this approach to treat two types of lysosomal storage disease in mice, suggesting that the approach could be used for other types of lysosomal storage disease and possibly other neurological disorders.

In addition to Davidson, the research team included UI postdoctoral researcher Yong Hong Chen, Ph.D., and Michael Chang, M.D., Ph.D., who was a student in the UI Medical Scientist Training Program when the study was conducted.

The study was funded by grants from the National Institutes of Health and from the Batten Disease Support and Research Association.

## Don't Forget -- You can now follow Dr. Jonathon Cooper and the PSDL on Facebook and Twitter

~ On Facebook, go to: <http://www.facebook.com/group.php?gid=131604681678>

~ On Twitter, go to: [http://twitter.com/Batten\\_PSDL](http://twitter.com/Batten_PSDL)

~ PSDL homepage, go to: <http://www.iop.kcl.ac.uk/departments/?locator=382>

## Upcoming Fundraisers

### Dart Tournament

Becky Lucas, mom to Chris Gaines (LINCL) is hosting a dart tournament on January 23, 2010. The event is being held at the Roebuck Elks Lodge in Birmingham, AL. We wish Becky and the Elks Lodge the best of luck!

### 4<sup>th</sup> Annual OurBoys 5K

The 4th annual ourboys 5k will be held on Saturday May 8th, 2010 at Harris Road Middle School, please save the date. 8am 5k and 9am One mile fun run. Online registration is open at: <http://www.sportoften.com/events/eventDetails.cfm?pEventId=4902>. 5k is \$25.00, Family of four is \$60.00 and 1 mile fun run is still \$10.00. Please be aware that this year the processing fee is included in your pricing. Please tell your friends and pass this around. We are still seeking sponsors, so don't pass up the opportunity to advertise your business and support a great cause!

### Battin' for Batten Disease First Annual Softball Tournament

The Joyce family of Blue Springs, MO will be hosting their softball tournament fundraiser on May 22, 2010. It will be held at the Blue Springs South High School. For more information, please contact Clarissa Joyce at [cjoyce1207@sbcglobal.net](mailto:cjoyce1207@sbcglobal.net).

### Paws for a Cause

This dog walk fundraiser will be held May 22, 2010 in Gahanna, Ohio and is being hosted by the BDSRA National Office. Dr. Martin Katz will be a guest speaker and will talk about Batten Disease as well as dog breeds that are predisposed to inheriting the disease. For more information please contact Adina Ryan at [aryan@bdsra.org](mailto:aryan@bdsra.org).

## *International Special Child - Rhiannon Bates Late Infantile Batten Disease - Derry, Ireland*

We were so proud when we finally became parents to a perfect little girl in June 2000. We were a very happy family, and we took the time to enjoy every moment of parenthood. As soon as mum was fit I went back to work, and mum stayed looking after our bundle of trouble. One thing mum did notice was that Rhiannon seemed to be having minor absences. We didn't recognise them as seizures at that time but whilst playing Rhiannon would suddenly pause, just like pausing a DVD or video, her lips and fingertips would turn blue, and she would stay like that for 30 seconds or so then continue on as if nothing had ever happened. As I was working I didn't get to see any of these for some time, but mum was worried enough that we got Rhiannon seen by a paediatrician, and they ran a heart tape and did some other basic tests but couldn't find anything wrong. After a few months these episodes seemed to die down, so we put it down to a phase Rhiannon went through and carried on with our lives.

All of this changed on Jan 1, 2004. Rhiannon was just 3 1/2 when she had an absence seizure in the bath. The local hospital wrote it off as an accidental near drowning, but we weren't convinced. There was no water in her lungs and no sign of her having swallowed any, either. Several weeks later Rhiannon had her first major convulsive seizure and was diagnosed as epileptic and started on an anti-convulsant.

Things started to go downhill from there. Within a year Rhiannon was showing a huge loss in gross motor skills and was experiencing problems coping in a mainstream class. She was assessed as having moderate learning difficulties and assigned a one-to-one classroom assistant to help her cope. This didn't work out at all – within 4 months she was re-evaluated and this time was assessed as having severe learning difficulties and moved to a special needs school. There she thrived, loving every minute of it, and she still does to this day.

When Rhiannon first started at her school she was able to walk and even run down the corridors, but that didn't last more than a year or so as she continued to deteriorate at a frightening pace. Speech and sight also continued to get worse and worse. It's interesting to note that it was about this time that our local consultant eye specialist suggested to Rhiannon's neurologist that she be tested for Batten's disease due to the damage to her eyes; her neurologist said she didn't fit the disease's symptoms.

It wasn't until Rhiannon had undergone two years of extensive tests at our nearest children's hospital with no sign of a diagnosis that we requested a referral to Great Ormond Street. We were seen within 3 months and left with a diagnosis of Late Infantile Batten Disease. At this time we were also informed that Rhiannon had now lost all of her sight.

One of our big saviours has been the Ketogenic Diet. After trying several combinations of anti-convulsant medication (which did everything from nothing to turning our child into a screaming monster), we put our foot down with her neurologist and insisted that she be started on the Ketogenic Diet. The results were obvious within a week or so; we had our little girl back and mainly seizure-free. The diet hasn't quite kept pace with Rhiannon's deterioration, so she has some medication as well now, but it has given us so much more quality time with her.

These days Rhiannon likes nothing better than getting mucky, be it with mummy making things like Christmas decorations out of clay or with daddy baking fresh bread and buns. We are also lucky to have a great family, and Rhiannon loves to sit on the sofa amongst us listening to all the gossip. As mum plays the trombone, music has always played a massive part in Rhiannon's life. The Ulster Orchestra has invited her to several concerts as their guest of honour, and she loves being the centre of attention.

We have always fostered the ideal that whilst Rhiannon may have a disability, it doesn't have her. She has travelled all over America (twice!) and across a good part of Europe. Even now in the advanced stages of Batten Disease we still manage to get Rhiannon abroad at least once a year for a holiday and a chance to recharge our batteries. She leads a very full and active life. She has her bad days and days where she needs a lot of sleep, but I think she is giving us a chance to rest more than needing it herself!

All in all we have been blessed with an amazing child. We won't have her forever, but the time we have with her is so precious. We take each day as it comes and make the most out of them that we can.

Written by her loving parents, Alan and Shauna

## Changes and Additions to the BDSRA Website

If you haven't visited the BDSRA website lately, we urge you to visit the site and take a look. Some of our newest additions include a fully integrated Bulletin Board. The Bulletin Board is very similar to the original style found on our old BDSRA website. We have also added a BDSRA store! Now you can purchase BDSRA items like our Batten Bears and BDSRA magnets right through our website. There are more great things to come so be sure to check back often.

## Garrett's Wings and Hayden's Hope Challenge Grant

Two \$25,000 challenge grants have been offered to BDSRA to help fight Infantile Batten disease. Garrett's Wings and Hayden's Hope have each committed up to \$25,000 for those making contributions to infantile Batten disease research before February 1, 2010. Last year, BDSRA committed \$194,000 toward this research effort and to date have raised over 50% of the funds. These grant opportunities, coupled with your donations, gives us a chance to meet this goal and raise \$100,000 for this vital research project.

## www.FightBatten.com

Josh Smerdel is a father, artist and advocate. His son, Mason, was diagnosed with JNCL in 2008. He has created a website to sell his amazing t-shirt designs and help build awareness for Batten Disease. His items are available for purchase now. You can visit Josh's website and view his items at <http://www.fightbatten.com>. You can place your order by emailing josh at [josh@fightbatten.com](mailto:josh@fightbatten.com).



## Capital One Card Lab Connect Benefits BDSRA

Share your passion for BDSRA and donate to our cause with your everyday purchases. We've recently partnered with Capital One Card Lab Connect to bring you our newest fundraising program, which helps us earn money effortlessly every day! Just carry one of our custom credit cards and 1% of your purchases made with the card will be donated to our organization. We will also receive a \$25 bonus donation when you make your first purchase. Not only will you be donating to our cause with each purchase you make, but you will be helping to spread the word when people see your unique card, designed specifically for our organization.

Visit our webpage: [www.cardlabconnect.com/bdsrasupportresearch](http://www.cardlabconnect.com/bdsrasupportresearch) to apply today and participate in this program!

## BDSRA Medical Equipment Exchange

BDSRA maintains a Medical Equipment Exchange for its families. We have many pieces of equipment available including wheelchairs, suction machines, bath aides, feeding supplies, etc. These items are available to BDSRA families at NO COST! If you need a piece of equipment or have equipment to donate, please contact Amy Lombardi ([kirka@bdsra.org](mailto:kirka@bdsra.org)).

## 2nd Annual Hayden's Hope Harley Ride

The Harley Ride will take place Saturday, May 22, 2010. The ride starts at Stocks Harley Davidson in Manitowoc and ends at the Iron Buffalo Salon in Menchalville. For more information please visit the Hayden's Batten Disease Foundation website at: <http://www.helphayden.com>.

## 3rd Annual Hayden's Hope Benefit

The benefit will be held Saturday, May 29, 2010 at the Two Rivers Community House. Activities will include a Kid's Carnival, auction, food & beverages. For more information, please visit the Hayden's Batten Disease Foundation website at: <http://www.helphayden.com>

## Nick's Golf Outing

A golf outing in honor of Nicholas Wellner is in the works for June 5, 2010. The event will take place near Nick's home of Waterloo, IA. More information to come.

## GoodSearch and GoodShop

You can use GoodShop to buy your favorite gifts and everyday items and earn money for BDSRA at the same time. Visit [www.goodshop.com](http://www.goodshop.com), designate BDSRA as your charity and choose the store you wish to shop. A portion of your total purchase comes back to BDSRA. We have raised over \$1300 this year thanks to you. Please visit [www.goodsearch.com](http://www.goodsearch.com) and designate BDSRA as your charity. Every time you search the web, BDSRA earns one cent! As you can see, these pennies add up very quickly! **Spread the word!**

## Family Services News

Amy K. Lombardi, MSW, LSW  
Coordinator of Family Services

### The Importance of a Seizure Emergency Plan

Even patients with well-controlled seizures have breakthrough seizures, and seizures can happen anywhere and at any time. Of the 67 percent of schools that call emergency medical services (EMS) annually for a student, 16 percent of these calls will be for seizures. One of nine schools can expect to have a seizure emergency at their school annually. Because this is such a prevalent issue, the benefits of an emergency treatment plan must be stressed. As Kathryn A. O'Hara, RN, an Epilepsy Nurse Clinician and Nurse Manager in child neurology explains, "Anyone, even those people well controlled, are at risk for a seizure so a plan should be in place as to what to do if a seizure occurs. The plan should include:

- a description of the person's seizure(s)
- how long it lasts
- what the postictal period (the time frame following a seizure) is like

It's important to know the difference of what is typical for a patient and what is not." A well executed plan reduces morbidity and mortality, empowers school personnel to respond to a seizure emergency, potentially prevents emergency department visits, helps reduce negative social consequences, minimizes classroom disruption, and reduces school liability risks. The goal of having the proper plan in place is to stop seizures quickly, remain safe in the school setting while experiencing a seizure, recover satisfactorily after the seizure, and return to the classroom as soon as possible. According to Orrin Devinsky, MD, who is Professor of Neurology, Neurosurgery, and Psychiatry at NYU School of Medicine and directs the NYU Comprehensive Epilepsy Center and the Saint Barnabas Institute of Neurology and Neurosurgery, "All children with active epilepsy should have an emergency plan in the school environment to make sure that the right first aid measures are employed and

that potentially dangerous interventions are avoided. Teachers, school nurses, and others should be aware of the child's seizure type and their typical duration. They should be familiar with first aid, including when to call for help or administer rescue medication." A solid plan begins with the basics: the student's name, the parent and physician's contact information, the type and duration of the seizure that might be expected from the particular student, and the current medications and dosages that the student uses as well as whether or not that medication is available at school. All activity involved with the seizure should then be carefully documented.

### Training School Personnel

An integral part of any seizure emergency plan is the training and education of school personnel. School personnel should be taught to:

- Recognize seizure activity
- Understand causes, triggers, and presentations specific to any given student
- Differentiate between what is typical seizure activity and what is atypical, potentially more serious, activity specific for an individual student
- Administer first aid for seizures
- Know when to call for caregiver(s) responsible for managing seizure emergency
- Know appropriate postictal care
- Know when to call EMS

Many school districts require training in regards to seizures and first aid, but many do not. The school nurse can provide training or the school can contact their local Epilepsy Foundation and ask for training, and many Epilepsy Centers offer training seminars.

BDSRA has a sample plan available for parents to view and adapt. It was written by one of our BDSRA fathers. It is important for schools to know what the parents' requirements are for treating their child's seizures. Do you want the EMS called if your child has a seizure? Do you have medicine available at school for emergency seizure situations? Should your child be allowed to rest after a seizure? These are all important aspects to consider when writing your plan.

You can receive a copy of the plan by emailing Amy Lombardi at [kirka@bdsra.org](mailto:kirka@bdsra.org) or calling her at (888) 379-2546.

(Article sampled from: "Planning Ahead Can Save the Life of a Child with Epilepsy" *By Laura Apel and Jan Carter Hollingsworth*. The entire article can be viewed by going to: <http://www.eparent.com/uploads/1/WEB-ed-epilepsyvaleant.pdf>)

### Faith's Lodge

The mission of Faith's Lodge is to provide a place where parents and families facing the serious illness or loss of a child can retreat to reflect on the past, renew strength for the present and build hope for the future. The lodge is designed as a place of respite and retreat. Families invited to come to the lodge include those who have children ages 1 to 19 facing a terminal illness and parents who have lost a child between the ages of 1 to 19 within the last three years. Families and parents are invited to stay between one and five nights. Faith's Lodge asks only that families give a \$25 donation for each night they stay. Located on 80 picturesque acres in the North Woods of Wisconsin, Faith's Lodge has eight individually designed guest suites that can each accommodate up to six people. For more information, visit their website, <http://www.faithslodge.org>.

### Camp New Hope

Located in the beautiful mountains of Ashe County near Boone, NC, Camp New Hope is a **privately owned, nonprofit, no-charge facility** for families who have children with life-threatening medical conditions. This unique and **rustic five-bedroom lodge** will provide you and your family with all the comforts of home, and more. Camp New Hope has been visited by many BDSRA families. Your only requirements to visit Camp New Hope are clothing and a toothbrush. Randy and the wonderful staff take care of everything else. This is the perfect getaway for any family facing the trials of Batten Disease. For more information, visit the website at <http://www.campnewhopenc.org>.

## **From the Nurse's Corner**

by Nancy Carney  
[nancycarney@bdsra.org](mailto:nancycarney@bdsra.org)  
1-877-642-5512

The next several issues of the Illuminator from my standpoint will be considering children with Batten Disease having delirium and acute confusion versus dementia. Since we are limited in the space we have to write our sections, I feel that the Delirium one will probably take at least two issues of the Illuminator as well as Dementia will also take at least another two issues of the Illuminator. I have written a book on Dementia and my information will come from there. If anyone would like a copy of that, you can email me or call me and I can send you one free of charge.

### **Delirium and Acute Confusion**

Patients in palliative care likely are experiencing severe exacerbations of chronic illness and use multiple medications - which put them at risk for changes in mental and cognitive function. Frequently, the new onset of behavior labeled as "confusion" is indicative of the acute syndrome of delirium. Delirium is a serious neuropsychiatric complication that unlike dementia is potentially reversible. It must be properly diagnosed and promptly treated in the palliative setting.

Descriptions of behaviors commonly noted in delirious patients can be found in medical writings from the time of Hippocrates to the present.

Despite this history, terminology has been inconsistent, overlapping, poorly defined and often adapted to the discipline or specialty observing the condition. Historically, terms such as organic brain syndrome, acute secondary psychosis, and sundown syndrome have been used synonymously with delirium. A review of the recent literature reveals a continuing use of a variety of terms to characterize delirium, including acute brain failure, acute confusional state, terminal restlessness, or agitation and psychosis. The most recent diagnostic criteria for delirium are as follows:

- Disturbance of consciousness with reduced ability to focus, sustain or shift attention.
- Change in cognition or the development of a perceptual disturbance that is not better accounted for by a preexisting, established, or evolving dementia.
- Disturbance develops in a short period of time and fluctuates over the course of the day.
- Evidence from history, physical examination, or laboratory findings that the disturbance:
  1. Is the psychological consequence of a general medical condition?
  2. Developed during substance intoxication or medication use.
  3. Developed during or shortly after a withdrawal syndrome.
  4. Has more than one etiology (e.g. more than one medical condition or a general medical condition plus substance intoxication or medication side effect).

Based on this criteria, delirium may be defined as an acute and fluctuating organic brain syndrome characterized by global cerebral dysfunction that includes disturbances in attention, level of consciousness, and basic cognitive functions (thinking, perception, and memory). Other features commonly associated with delirium include increased or decreased psychomotor activity, disturbances in the sleep-wake cycle, and emotional lability (being unstable or changeable). Delirium is frequently unrecognized by clinicians or misdiagnosed. The fact that demented, depressed, and anxious patients may develop delirium makes the diagnosis additionally difficult. The diagnosis of delirium is primarily clinical and requires careful observation and a thorough history. Because the signs and symptoms of delirium are nonspecific, the clinician must look for a constellation of findings, identify the rapidity of onset, and assess for associated medical and environmental risks to determine an appropriate diagnosis.

Key features of delirium are:

1. **Disturbances of consciousness** which refers to impairments in attention and the ability to be aware of and sustain to the environment. Attention is typically fluctuating and may present as a change in the level of consciousness that does not reach the level of stupor or coma. Patients may demonstrate slowed or inadequate reactions to stimuli or manifest distractibility.

*Continued on page 7*

Individuals may be unable to follow conversations or complete simple tasks, may have slow response time, may be unable to maintain eye contact, or may fall asleep between stimuli. Increasing stimuli (touch, sound) may be needed to elicit a response. Conversely, patients may be hyperalert and over attentive to cues and objects in the environment. The ability to focus can be assessed by the patient's ability to complete a particular task such as spelling the word backward, subtracting serial 7s, or listing the days of the week in reverse order.

**2. Change in Cognition** - many aspects of cognitive function are impaired in delirium, including orientation, memory, language, thinking, and perception. Disorientation usually manifests relevant to time or place, with time disorientation being the first to be affected. Disorientation to other persons occurs commonly, but disorientation to self is very rare. Short term memory deficits are the most evident memory impairments. Immediate memory loss (over a period of seconds) is demonstrated by the digit span while memory (over a period of minutes) is demonstrated by the ability to remember three objects after 5 to 10 minutes. However, because these tests also reflect attention, severe deficits in this area will result. Language disturbances include a lack of fluency and spontaneity (long pauses and use of repetitious phrases), a tendency to ramble and switch from

topic to topic, and difficulty finding the correct word to use in conversation or naming objects. Thinking is usually disorganized as evidenced by incoherent speech, deficits in logic, and responses that are irrelevant to questions asked. Perceptual disturbances may include misinterpretations, illusions, or hallucinations.

Visual misperceptions and hallucinations are most common, but auditory, tactile, gustatory (sense of taste), and olfactory, (sense of smell), misperceptions or hallucinations can also occur. The individual with delirium may have the delusional conviction that the hallucination is real and exhibit emotional and behavioral responses consistent with the hallucination's content.

**3. Acute onset and fluctuating course** - the features of confusion that develops over a short period and fluctuates over the course of the day are important in defining criteria for delirium. Symptoms develop over hours or days but are likely to be intermittent in presentation and severity. A typical presentation is worsening of symptoms at night with lucid periods during the day where the patient may function normally. Noting the onset of the disturbances in consciousness or cognition assists in differentiating delirium from other syndromes that cause mental status changes, such as dementia.

**4. Etiological Evidence** - An important criterion for the diagnosis of delirium is evidence that the changes are a physiological consequence of an

underlying medical condition, substance or medication intoxication or withdrawal, or combination of these factors. In palliative and end of life care, it may be difficult to identify the exact cause of the delirium. An individual may have several potential causes at any one time. The challenge for the clinician is to identify which of the potential causes is the most likely and then determine the appropriate approach to this problem.

**5. Sleep-wake disturbances** - in sleep patterns include daytime sleepiness. Nocturnal insomnia, disturbed sleep continuity, and excessive dreaming. Some patients may experience a complete reversal of the sleep-wake cycle characterized by diurnal (happening in the daytime) sleep periods and nighttime agitation and insomnia, whereas others may have fragmentation of the circadian sleep-wake characterized by short periods of sleep and waking across the 24-hour day.

**6. Psychomotor Activity** - Patients with delirium may also exhibit disturbed psychomotor activity. Continuing research has indicated that there are several subtypes of delirium based on motor activity. These include a hyperactive-hyperalert subtype, a hypoactive-hypoalert subtype and a mixed subtype that features components of the other two. To briefly share with you the difference of these subtypes, the first one shows evidence of sympathetic nervous system over activity manifested in restlessness or agitation or inappropriate behavior.

## In the Works....

Adina Ryan, Director of Development

Email: [aryan@bdsra.org](mailto:aryan@bdsra.org)

Phone: 1-866-287-7233

## Happy New Year everyone and welcome to all our new families!

As we begin 2010, we need to celebrate our achievements and all that is about to come. As we look forward, we must keep our sights high and focus not only on the past year's enormous challenges, but also on the accomplishments we have achieved together. Since my arrival here in mid October, I have witnessed a great deal of accomplishments for 2009 including the fulfillment of our financial commitment of \$400,000 to the Juvenile drug trial, providing support services this year to more than 120 families (60 new families), a first ever annual fund drive totaling more than \$47,000 (includes a major gift of \$30,000, and an updated website design. Just to name a few.

*Thank you for your support in my new role here at BDSRA and for your generous contributions. Your gifts are supporting the mission, programs and services of BDSRA so that our staff can continue to provide you with all*

*the information and materials you need, and supporting the researchers in their efforts to find treatments and a potential cure for our children.*

## Use Your Will to Change the Future

**Want to change the future for children diagnosed with Batten disease?** A bequest in your will allows you to help change the lives of children tomorrow without parting with any of your assets today. Plus, your generosity will continue long after you're gone. There are two special features:

### 1. It gives you some flexibility.

A bequest lets you balance your philanthropic goals with your concerns about your future. Because you're not actually parting with assets today, you don't need to worry that you might later need those assets to live on. Plus, you can change your mind at any time.

**2. It lets you be sensible in your commitment.** Your gift can be made as a percentage of your estate, allowing you to benefit your favorite causes and loved ones in relative proportion.

## How to Make a Gift in Your Will

- Decide what amount or percentage you want to give. A percentage gift ensures that the size of your gift will remain proportionate to the size of your estate, no matter how it fluctuates over the years.
- Take our official bequest language (provided on request) to your estate planning attorney to add to your will.

- Work with your estate planning attorney to update your existing estate plan with an addition or to begin developing your estate plan.
- Notify us of your intention so our staff can thank you for your future gift and keep you informed of our ongoing activities. (*we will honor your preferences regarding anonymity*)

## Questions?

Feel free to contact Adina Ryan, Director of Development at 866-287-7233 or via email at [aryan@bdsra.org](mailto:aryan@bdsra.org)

## Are you having an upcoming fundraiser?

Please let BDSRA know if you are planning a fundraiser. We can send you brochures and other materials to help promote your event and educate your attendees. BDSRA merchandise is also available for purchase for you to resale at your event. Please contact Adina today to let her know about your amazing upcoming event.

## 2009 Audit and 2009 Annual Report now available on BDSRA website

Are you interested to know where your donations are being used? Want to see what has happened in BDSRA over the past year? Please check out the 2009 Annual Report and Audit on the BDSRA website: <http://www.bdsra.org/about.html>



## Fourth Quarter Honor Roll

*BDSRA has been remembered many times in the past three months by family and friends of children with Batten Disease. To all of you we express our deepest appreciation.*

### **MEMORY OF ZACHARY AHLSON**

Esty Optical Company

### **HONOR OF DEREK ALLBEE**

Cal Neva 7 Eleven Franchise Owners Assoc

### **HONOR OF THE ALLIO FAMILY**

FPOA Batten Disease

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### **HONOR OF TYLER ALLMAN**

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Ms. Shelly Rice

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Mr. & Mrs. Dean Bucalos

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H. Degraw

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Head and Heart Coaching

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Ms. Florence Petri

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*BDSRA has been remembered many times in the past three months by family and friends of children with Batten Disease. To all of you we express our deepest appreciation.*

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Ms. Susan Converse

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Ms. Diane Adams

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Mr. & Mrs. Kevin Reilly

Mr. & Mrs. Jerry Daily

Ms. Mary Mero

Ms. Janet Oley

Mr. & Mrs. Stephen Healy

Mr. & Mrs. Reginald Adams

Ms. Suzette Pfiefer

Mr. & Mrs. George Adams

Mr. & Mrs. George Hay

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Mr. & Mrs. Craig Montgomery

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Mr. Brock Frericks

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Mr. Don Ferman

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### **MEMORY OF ELAINE, LEE, &**

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### **MEMORY OF ELAINE JOHNSTON**

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Ms. Kathleen McGuigan

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Xinhai Ni

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### MEMORY OF ELIZABETH ANN DAVIS

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#### 31<sup>ST</sup> BIRTHDAY

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Mr. & Mrs. Michael Guertner

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Mr. & Mrs. Mike Dodge

### MEMORY OF KAREN ELISE NEWTON

Mr. & Mrs. Robert Newton

### MEMORY OF MEGAN NICOLE

#### DEPEW

Phyllis Depew

### MEMORY OF ABBY BORTZ

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Mr. & Mrs. Gregg Froio

### MEMORY OF LAUREN RANKIN

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Mr. & Mrs. Jack Caveney/ on behalf

Midwest Chptr

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W.L. Haddad

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#### BECKY GJEMSE

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Lamira Jaecks

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Mr. & Mrs. Jim Tobalski

Anonymous

Ms. Gloria Rowan

Ms. Lottie Sutton

Ms. Sandra Sellan

Anonymous

Anonymous

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Mr. & Mrs. C. Thomas Fennimore

Mr. & Mrs. Kurt Moser

Mr. & Mrs. Wasyl Karpenko

Ms. Kathlyn Hames

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Noah's Hope Fund of

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Mr. David Pasnes

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### HONOR OF CELIA BETZ

Mr. & Mrs. James Betz

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Mr. & Mrs. Warren Shuros

Mr. & Mrs. Michael Neal

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Ms. Mary Giustino

Mr. & Mrs. Francis Casta

Mr. & Mrs. Thomas Stone

Ms. Holly Hawkins

Mr. & Mrs. Eric Faret

Mr. & Mrs. Jeffrey Bolick

Marian Dealey

Mr. & Mrs. Gary Thomas

#### Juvenile Trial

Alpha Chi of Alpha Delta Kappa

Lisa Gilbert Photography

Gervaise Designs

The DuPage Community Foundation

As Suggested by The Van Houtan Family

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Mr. James Montville

Kathryn Bourque

Mr. & Mrs. Mark Ducharme

Mr. & Mrs. Dennis Cilley

Mr. Kyle & James Pelletier

#### STIPEND

Anonymous



# *In Loving Memory*



**Tom Smyser**, husband of Tracy and father of Shaun (deceased) Smyser, Stockton, IA  
Died: 9/14/09

**Jamie Mitzel**, daughter of Roger and Sheri Mitzel, McCoy, TX  
Born: 02/23/89 - Died: 10/08/09 Juvenile NCL

**Blake Hux**, son of Hunter and Heather Hux, Mendenhall, MS  
Born: 04/10/98 - Died: 10/10/09 Unknown NCL

**Josee Pitchette**, daughter of Gilles Pitchette and Jacqueline Violette, Grand Falls, NB Canada  
Born: 03/28/78 - Died: 10/17/09 Infantile NCL w/ Juvenile Presentation

**David Pfohl**, son of Warren and Brenda Pfohl, Manlius, NY  
Born: 11/02/88 - Died: 10/22/09 Infantile NCL w/ Juvenile Presentation

**Brandon Smith**, son of Doug and Cindy Smith, Winnipeg, MB Canada  
Born: 03/06/01 - Died: 11/01/09 Late Infantile NCL

**Chelsea RaNae Garza**, daughter of Alex and Terri Garza, San Antonio, TX  
Born: 09/10/92 - Died: 11/20/09 Late Infantile NCL

**Zachary Ahlson**, son of Derek & Rachel Ahlson, Rhododendrun, OR  
Born: 01/20/03 - Died: 12/01/09 Infantile NCL

**Amanda Fitzgerald**, daughter of Donna Fitzgerald, Elgin, SC  
Born: 09/02/81 - Died: 12/05/09 Juvenile NCL

**Amber Royalty**, daughter of Randy and Darlene Royalty, Ursa, IL  
Born: 03/15/81 – Died: 12/17/09 Juvenile NCL

**Sarah Royalty**, daughter of Randy and Darlene Royalty, Ursa, IL  
Born: 10/05/85 – Died: 12/19/09 Juvenile NCL

**Aleah “Allie” Galen**, daughter of Ben and Melinda Galen, Somerset, WI  
Born: 05/31/97 – Died: 12/30/09 Juvenile NCL

**Adam Hawthorn**, son of Will and Pam Smith, Black Falls, AB Canada  
Born: 01/26/89 – Died: 01/06/10 Juvenile NCL

**Timothy Nordquist**, son of Don and Kathleen Nordquist, Bismark, ND  
Born: 02/02/83 – Died: 01/07/10 Juvenile NCL

**Hailey Goranflo**, daughter of Neil and Miranda Goranflo, Shepherdsville, KY  
Born: 10/30/02 – Died: 01/17/10 Late Infantile NCL