This Q & A article is from BDSRA’s “Ask An Expert” Facebook Chat in the BDSRA Closed Facebook group for parents and caregivers.

\*Please note: the information contained in this material is intended to provide

basic information to Batten families and caregivers. It is not intended to be, nor is it, medical advice for individual children. Parents and caregivers should consult the patient’s physician prior to changing medication, medical treatment

or daily activities.

“Managing Meltdowns and Tantrums”

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Introduction

The University of Rochester Batten Center is located in Rochester, New York. We have a very large team here, including the two of us on the call (Amy and Heather) and also pediatric neurologists Dr. Erika Augustine and Dr. Jonathan Mink. Dr. Mink is the Director of the URBC and both he and Dr. Augustine are experts in Batten disease. Also on our team are two outstanding research coordinators, Ms. Alyssa Thatcher and Ms. Sara Defendorf, the director of our genetic testing lab, Dr. Paul Rothberg, PhD, and Dr. Frederick Marshall, MD who is an adult neurologist. We also are proud to include many students on our team every year, including undergraduates, medical students, and graduate students.

Amy Vierhile, here on the Facebook Chat is a pediatric nurse practitioner and has worked in Child Neurology for the past 19 years. She also was instrumental in helping to execute our recent clinical trial of mycophenolate for JNCL, the “JUMP” trial. Heather Adams, also here on the Facebook Chat is a clinical child psychologist and pediatric neuropsychologist who has been practicing and doing research for about 15 years. She has published studies on the cognitive and behavioral aspects of Batten disease and also just published a paper on using telemedicine (video-based) assessment of cognitive abilities in JNCL.

To get conversation going…
We would like to ask a couple of questions to the Batten community, about challenging behaviors in children with Batten disease. What are the behaviors that cause the most difficulty for your affected child, you, and your family? And, thinking of those most challenging behaviors, what are some examples of how the behavior(s) impact your affected child and others?

Q:Hello, my daughter, has CLN3, she is 11. We have a lot of meltdowns from her getting fixated on things...wanting a specific toy that may be misplaced or a new toy. Some of our other meltdowns are focused around frustration. As far as the impact, I feel like we tend to walk on eggshells, avoiding some topics for fear of setting her off.

For example, if she's recently been fixated on Hello Kitty toys then we try not to talk about it, ESPECIALLY if one has been missing. The "stuck" part is tough....we might get her redirected briefly but it seems like we are right back to it within 1/2 an hour...and then there can be crying and yelling fits then can become physical - hitting or throwing things. She has a lot of perseverance in her speech and behaviors...so I think it is partially related to the memory/dementia issues.

A:Hi, thank you for writing in. Both of these issues are ones that we hear a lot about in children with JNCL/CLN3. They can indeed get very focused on certain topics, really getting 'stuck' like it's a broken record in a way. We also know that some children have a temperament that is designed that way, and some do not end up having the issue of getting stuck on ideas. Some of the 'getting stuck' might be due to forgetfulness - we know that JNCL causes problems with learning and memory, and that over time, children have a harder and harder time keeping track of what they have said or done.

Q: So besides redirection and avoidance, do you have any other suggestions?

A: We always want to put safety first. Redirection is a good strategy that you are using. One of the tricks to try is to redirect the child to an activity that is even more preferred than the thing they are getting stuck on. The other thing to try is to provide lots of attention and praise during those brief moments when they have been able to get 'un-stuck', and to try to stretch out those times. But I agree with you, you are absolutely right that dementia is contributing. Even with adults who have dementia, getting 'stuck' is a common theme. You might hear the docs referring to this as "perseveration". Amy Vierhile here is also commenting. She adds that some medications might help to lower aggression and perseveration by taking the edge off their anxiety and their reactive style. And (from Heather) the other thing to add is that it can be really really helpful to have a local behavioral specialist come to observe what is going on and perhaps do what is called a "functional behavioral assessment". I am going to add another post shortly to explain that. Becky from the BDSRA also reminded me that it is also important to think about how to support and explain this to siblings, who may be confused or frustrated themselves. Obviously, the explanation you give will depend on the age/maturity of the siblings.

A: Let me explain about a Functional Behavioral Assessment (FBA); sometimes, it can be helpful to do what is called a “Functional Behavioral Assessment” or “FBA” to better understand the factors that both influence a child’s behavior, and are influenced by a child’s behavior. An FBA uses a variety of techniques to understand what is happening before, during, and after a certain behavior occurs. The things that happen before might either trigger a behavior or might lower the chance of the behavior occurring. A s a very basic example, think about a child who usually is very happy to pick up their toys when asked, but occasionally has a tantrum when this request is made. Perhaps the triggers for the tantrum have to do with being tired or hungry, or feeling mad already because they just had an argument with a friend or sibling about something else. The FBA would help look at the patterns of these triggers, and then come up with suggestions for how to deal with them. In this example, perhaps one possible solution would be to make sure the child has a snack when they come home from school, before they are asked to clean up their toys. The other piece of the FBA is to look at how to what raises or lowers the chance of a behavior continuing (e.g., the “during” piece) and what happens after. For example, does talking to the child during their tantrum help, or does it add fuel to the fire? Every good parent’s first instinct is to step in when their child is struggling or upset. But during a tantrum, it might be helpful to let it run its course (assuming that there are no safety concerns) and then talk about it afterwards. The ‘after’ piece is important too – does the outcome or response make it more or less likely that the same behavior will happen again another time?

**There is a wonderful website that explains all about FBAs:**[**www.cecp.air.org/fba/**](http://www.cecp.air.org/fba/) **This is the Center for Effective Collaboration and Practice**

**P**lease encourage the specialist to contact the BDSRA to learn about Batten disease, and we here at the University of Rochester Batten Center are also always glad to talk about Batten disease if other specialists have questions. Amy fields these calls from providers all the time!

Q:Who would you work with to get a referral for a Functional Behavior Assessment (FBA)? Are they usually done in the home setting?

A:To address the question about getting a referral for FBA... I would start with the local school district, if your child is still enrolled in school. Otherwise, you could contact a local child mental health professional or, if you live near a hospital with a psychiatry department, you could contact that department. Go to the agencies and programs that serve individuals with developmental disabilities such as Autism or Down Syndrome. They have great expertise as FBAs.

Regarding where to do the FBA, it is often done in multiple settings, because children spend their time in multiple settings. Since your child is at a residential school, the FBA might need to be done there as well as at home. There might be different triggers or strategies to consider depending on the environment (school vs home). The other thing is that the School might need some specialized training in how to support a child who has dementia. Most schools are built on the model of children \*gaining\* skills over time, but with Battens we have two opposing tides of development and regression. It can be tough for some teachers to understand and accept that.

Q:If we do an FBA at both locations, would you need the same specialist to complete the observation? (we live 5 hours from the State School for the Blind). If we do an FBA at both locations, would we need the same specialist to complete the observation? (we live 5 hours from TSB)?

A:I have to admit that I am not sure whether the FBA would be done by the same specialist at both locations. Ideally it would be, but I recognize the limitations due to distance between your home and school. I think it would depend upon first talking with some FBA specialists in your region to see how they would want to proceed

Q:What medications typically help with this kind of thing? She is not taking much right now. Just clonidine and her seizure med....ugh drawing a blank...zonegran.

A:Also, to address about medications that can help with challenging behaviors: Amy mentions that SSRIs, which are serotonin-specific reuptake inhibitors, are common ones for helping with mood and behavior. The other medication category would be what are called 'neuroleptics' (historically called antipsychotics). These are stronger medications but work better for aggression. ANY medication decisions should be taken with your medical team and the addition of a behavior specialist is always helpful - it's not just a medication OR behavior management choice - both are often needed.

Q:We are having problems with our 11 year old son, JNCL. He is having screaming fits for hours on end and nothing we do helps out. He ends up hoarse, sweating and exhausted. Transitioning from home to school and back is very difficult. Neurologist is unhelpful with ideas and I am at a loss. Also trying to get info if CBD oil can help behaviors?

A:We are sorry to hear what a tough time your son is having. Amy is going to address the question about CBD (Cannabidiol) oil. CBD oil is the oil from a cannabis (marijuana) plant but that does not contain THC. THC is the ingredient that makes people 'high'. So the CBD oil does NOT make your child high. Right now, there are clinical trials underway to determine if CBD oil is helpful for seizure control, but as far as we know, it is not being studied for behavioral control. Transitions are very, very difficult for children with dementia. They can be confusing to them, even for transitions that happen every day. In terms of behavioral strategies, creating transitions that build in enough time and are slow enough and gradual enough that the child almost doesn't know it's a transition, can be one trick. But this has to be planned out ahead of time, discussed with the whole team, almost scripted out.

Q: How can I determine dementia from behavior? I am assuming that is what this is as he constantly says Mom over and over and will ask, can I ask you a question literally 15 times in a row before he moves on. He gets very frustrated and lashes out verbally.

A:To address the question about dementia vs. behavior -- we view the challenging behaviors as one of the many symptoms of dementia. We have a saying at our clinic, "Children do well if they can" and I will post more about that shortly.

**Children Do Well If They Can**
We firmly believe that children do well if they can. That is, if a child has the skills and knowledge to do a certain task or behave a certain way, they will do so. Of course, children always have some skills that they have completely mastered, some that they are working on learning, and some they are not yet ready to try. We can see behavior issues in the zone where children are either asked to perform a skill that they are still working on (so perhaps they can only do it sometimes, or in certain situations), or when they are asked to use a skill that is too hard for them. With Batten disease, this is very tricky, because children are both gaining some skills and possibly losing ground on others, at the same time. That is, there are some skills that they used to be able to do very consistently (such as…pay attention, pick up toys, handle frustration, wait their turn, etc.) that it becomes hard to do. If conditions are absolutely optimal they might still be able to use the skill. But if not (feeling tired, hungry, had a seizure that day, upset about something, in a new environment, etc), then it might be harder for them.

Skills can be things like the “how to” stuff (like, how to tie your shoes, how to throw a ball”) or they can be the “process” stuff (such as: waiting your turn, sharing toys, keeping hands to self). If a child interrupts us when we’re on the phone, it does not help us fix it by just labeling the child as misbehaving, or giving them a punishment. But if we reframe it as, “this child lacks the skill to wait his or her turn” then we can think about ways to either teach the skill if it can be learned, or accommodate the absence of the skill or the loss of the skill if it cannot.

Getting back to the discussion about behavioral management, we do want to emphasize that for individuals with dementia (children or adults), **structure and routine** are very, very important. Sometimes there are school environments that are more predictable or structured that are helpful, including residential schools that specialize in serving children with disabilities.

Q:I think structure may be part of our home issues. What things help to get more structure in place? How do you implement structure when they change their mind every 5 minutes about what they want?

A:That is an EXCELLENT question (about the kids changing their minds every 5 minutes). We admit, we struggle with this too! We know that how the adults approach the situation can help. Even though it is terribly frustrating, we know that staying calm helps a lot, if only for your own peace of mind. But also, kids with dementia are very sensitive to their environment, so when things are busy and stressful around them, they might also feel a bit more stressed themselves. They may not have good 'filters' for tuning out what is going on with other people.

Q:In your experience, do noisy environments tend to bother JNCL children? For instance, we are big football fans and love watching the games on TV on Saturday.

A:To your question about noisy environments, we think that each kid's triggers will be different. Some kids really enjoy a noisy busy environment, and other kids might prefer a quiet one. If a child is overwhelmed by a noisy environment, perhaps it would help to allow the child to wear noise-cancelling headphones and listen to some music or their favorite movie while others watch the game?

Q:We have a lot of issues with waiting her turn. She used to be able to do this, but now...not so much except on a really good day/time when she seems to have a lot of clarity.

A: Yes, exactly! On her good days, she can still use the skills. I know this can be so frustrating for parents, sibs, other family, and teachers - children can sometimes use a skill, and other times they can't. It can be very confusing - good day/time when she seems to have a lot of clarity.

Q:Our son wants to be independent but seems to have lost that ability so he needs us for everything and if we can't do everything immediately it can go to hour long meltdowns. Also have a 13 year old unaffected daughter that we worry about the impact on her. I can't find the balance. The balance is so hard! Our 13 yr old unaffected sibling also has trouble because of our daughter's temper, which is sometimes directed at her sibling, and her lack of patience/turn taking. It really frustrates the older one because she ends up waiting so much and having to "walk on eggshells" with her sister.

A:Yes, it is definitely really tough when children still remember the things they could do, and still can (given enough of a long time) but you need to step in and help. We (both of us) can't emphasize enough how important it is to have the behavior specialist involved to help come up with an action plan for when these issues come up. Finding things that kids can still do themselves (even the smallest thing) to help them feel they are helping, might be something to try.

To pick up on the other comment about your 13 year old sib, we would definitely want to make sure that she has someone to talk to about all this, like a counselor at school or in the community. It is really a tough position for them to be in, because so much of this is not in your control. There might be some things that could be done to help the child with Batten disease respond more positively to a sib, like providing a preferred activity to the affected child in the presence of the sib, or having the sib be involved in actually providing that activity or treat themselves. However, the safety should always come first, and it would be helpful to have a child behavior specialist involved with planning any of these strategies so they are done in the safest way possible.

Q:Our unaffected daughter clams up with counselors both school and private. We are having a horrible time getting her to talk to anyone about her frustration, anxiety and depression over being faced with something like this. She feels that none of her friends really can relate since they have not experienced anticipatory grief like this....we are worried about her.

Q:Same problem with our daughter. Getting better with seeing a counselor. The BDSRA conference was super overwhelming for her. Only 30 days diagnosed and we were there. She hid in the room most of the time. We are super worried about her too.

A:  I am so sorry to hear about this. I know it is heartbreaking for you. And I can absolutely respect a teen's reluctance to open up to people who might not understand. There are some options to consider that might help teens connect to others who do 'get it'. First, the BDSRA can help connect teens to one another. I know there is a sibs group and some of the older ones in that group might be good mentors for her. There may also be other groups in your community to support sibs of children with other types of serious or life-threatening illnesses (such as pediatric cancer). These kids won't know exactly what the Batten sib is dealing with, but can relate to the fears and stresses of having a brother or sister who has a serious illness.

Q:Agreed, our daughter is part of the sib group and a few have really reached out to her. I think she is looking forward to St. Louis now!

Q:We are pretty rural here but there is a Children's Hospital...would they be a good place to contact to try to find out about community support groups? Or are there other places to approach to link into support groups?

A:We do not have suggestions at my fingertips for specific places to go, but in the next day or so we will provide the BDSRA with some resources. BDSRA’s staff can help you locate a group in your area, if one is available. There are national and statewide organizations that help families of children with disabilities, and we will put together some resources that folks can investigate.

We want to now address the comment about discussing medications with her doctor. Here at the URBC we are more than happy to discuss medication issues with your child's doctor. Please contact us to discuss how to coordinate a conversation on this. Sometimes we can help a local provider become more comfortable with medications they don't usually prescribe; or, sometimes it helps to have a psychiatrist become involved to help with the medications for challenging behaviors.

 Q:I understand your concern about lack of knowledge/experience with Batten locally. That is part of the reason why we travel so far to see Dr Emily De Los Reyes at Nationwide Children’s Hospital in Columbus (8.5 hour drive one-way with our daughter). I have noticed your previous comments in other conversations about the lack of proactive care from your neurologist. Perhaps there might be someone else that would be willing to do their homework? Yes, the wonderful [Margie Frazier](https://www.facebook.com/margie.frazier.7?hc_location=ufi) and I will be speaking about that soon. He is just very dismissive and I get concerned when my son's neurologist tells me "he is not qualified to prescribe these types of meds". Our doctor did speak with Dr. Mink.

A:Your doctor who spoke with Dr. Mink, they are always welcome to contact us multiple times; they can check in with us whenever they want to. Becky also just reminded us to say that it may be hard when you are trying so many different medications - you might feel that your child is a bit of a 'guinea pig'. But every child is different and so there is no one specific magic medication that works in the same way for everyone. It is really important to try different combinations of medicines, but to do so in a thoughtful way.

Becky also asked us to address what types of challenging behaviors would lead to a more intensive level of care, such as an inpatient medical/psychiatric hospitalization. When the aggression reaches the point that the child cannot be safely managed at home/school, this might be a time to consider hospitalization. A hospitalization can be helpful for changing medications in a controlled setting to see if another medication strategy can help. It can also provide respite to the family while community supports are being put in place.

Q: Ok what would be an example of aggression that is out of control and not safely managed? We hospitalized our son in 2012 for aggression and they kept him for a week to tweak meds. I have been afraid to take him back because I am so afraid no one knows what to do.

A: To address your question of what is aggression that is out of control and not safely managed...it will depend on the child. Children who are small and not strong, but hit, may be safely managed at home. Children who are big and quite strong, and are hitting in a way that hurts others, may not be safely managed at home. However, we do recognize that hospitalization is a very hard thing to go through, and can be quite traumatic for everyone. We really view it as a 'last resort' choice, and we always prefer to solve these issues with children remaining in their home and community.

This brings us to another REALLY IMPORTANT point of making sure that as parents, you take care of yourselves!

We wanted to say thank you so much to the BDSRA for hosting this Facebook Chat topic, “Managing Meltdowns and Tantrums”. Thank you to everyone who posted questions and signed in to follow along. If we did not get to your question, or if there are additional questions later, please reach out to the BDSRA or contact us at the University of Rochester Batten Center. Our contact information is: [https://www.urmc.rochester.edu/.../batten-disease-center...](https://www.urmc.rochester.edu/neurology/batten-disease-center.aspx) and[https://www.facebook.com/URMCBattenCenter](https://www.facebook.com/URMCBattenCenter?hc_location=ufi)

The University of Rochester Batten Center website also has a free handout – based on a talk that Dr. Adams presented at the 2009 BDSRA Conference. The full link to the handout (a PDF of a powerpoint slide show) is:[https://www.urmc.rochester.edu/.../BDSRA2009...](https://www.urmc.rochester.edu/MediaLibraries/URMCMedia/neurology/documents/BDSRA2009_behavioralandeducationstrategies.pdf) but the easy way to find it is to just navigate to our main website page, click on the “Education” tab on the left-hand side of the page, and then download the PDF.

Finally, thank you especially to Becky Hetteberg, the BDSRA Family Support Officer, for organizing and helping to moderate this chat. On the website there is a link to our email as well.

[Becky Hetteberg](https://www.facebook.com/becky.hetteberg?fref=ufi), BDSRA:  Thanks very much to Dr. Adams, Amy Vierhile and Sara Defendorf sharing your expertise about behavior management with our Batten families tonight! We truly appreciate the fact that you have spent your evening with Batten families helping them learn more about this important aspect of care.

URBC:Thank you Becky! And thank you to all of the Batten families out there. We are so honored that you share your stories with us.