

CHARGE Accounts



Spring 2007

A Quarterly Newsletter for Families and Friends

Vol. 17 No. 1

CHARGE Syndrome:
CHARGE has four
major features -
Coloboma, Choanal atresia,
Cranial nerve abnormalities, and
Characteristic ears
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CALENDAR 2007

- June 15** **Deadline for Articles in the SUMMER Issue of CHARGE Accounts**
- June 30** **Summer Issue of CHARGE Accounts in the mail**
- July 27 - 29** **8th International CHARGE Syndrome Conference, Costa Mesa, California**
- September 1** **Deadline for Articles in the Fall Issue of CHARGE Accounts**
- September 25-30** **Deafblind International Fourteenth World Conference, Burswood International Resort Convention Centre, Perth, Western Australia.**
More details:
www.dbiconference2007.asn.au/
- September 30** **Fall Issue of CHARGE Accounts in the mail**



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!!! Deadline for the Summer Issue - June 1 !!!

Things are really moving along quickly as we approach the 4-month countdown to conference! Be sure to make your room reservations soon. Once our reserved block is full, the rates may change. We have reserved a few more rooms into the block, but those will probably disappear quickly. We are checking about being able to offer more rooms on the "shoulder dates" (prior to and after conference for those wanting to extend their time in California) at the reduced rate. Please be patient and understanding as we deal with the incredible response to bringing the conference west!

Childcare will be staffed by Kiddie Corp, a professional childcare service. If you have children who will be involved in childcare (Beach Club), you must be sure to register with Kiddie Corp. Details are in the registration package and on-line at www.kiddiecorp.com/chargekids.htm. We still request that you include the childcare info with your registration to the Foundation. This allows us to cross-check with Kiddie Corp that they have received the registration information they require. They want to do the best for us by having the appropriate staff numbers and skills. The only way to accomplish that is to have everyone directly registered with their agency.

We received great news that Variety Club has approved a grant to be used in areas pertaining to assisting the children to enjoy the conference. (Yeah, Ruth Schwartz! Great job getting that grant!) This will be used to enhance the fun experiences for our children. We will be bringing in crafts additional to those already planned for the Beach Club rooms. The staff from Michael's Crafts have some very cool projects planned! The Saturday evening event will also benefit from this grant.

We have also received funds from the Chores-4-Charity fundraiser started by Madison (the 10-year old sibling of an individual with CHARGE). This is an amazing project that you can find at www.chores-4-charity.com.

And don't forget about the Fundraiser-In-A-Box! Really easy! Really quick! Really needed! Contact the office for additional information if needed.

The California Deafblind Services staff is onboard to assist in suggesting any adaptations to activities, as well as hands-on assistance in the Beach Club area. They will also conduct the sensitivity training for the hotel staff. This is a great service for the Foundation and our conference attendees: To utilize the knowledge of the CADBS staff in this manner is to know that we will have good service and understanding from the hotel and staff. Jackie Kenley has been a great contact and help for me in many aspects of organizing the conference.

The design for the conference shirts and bags is completed and absolutely beautiful! You will be impressed and want one for every member of your family! Thanks go to Wendy Keedy for working with the graphic designer, and truly capturing California in the work.

Volunteers are still needed! The volunteer base needed to make the conference flow well is huge. It is also, by far, the most difficult task in organizing the conference! If you have connections (family, friends, friend-of-a-friend, religious organization...!) in the Costa Mesa/Orange County area, please contact and encourage them to get involved. In the past, we've had entire families volunteer to work and all have come away with a wonderful experience. We can utilize individuals 18 years and older for the direct childcare rooms, thus reducing the costs for the professional services. We also need volunteers for the craft area, and a large number for set-up/tear-down of the Saturday night event. These volunteers can be of any ages that are willing to contribute their time and energy. Scouting groups are a great resource for craft volunteers.

Silent Auction and Donations

Once again, we will have a silent auction during the International CHARGE Syndrome Conference. You should have already received the request letter and donation form in the conference registration package. Please ask for donations from groups or individuals who might have a product we could auction. Nearly anything is acceptable. In the past, we have auctioned resort weekends, hotel certificates, rugs, handmade items, rings/jewelry, chain store gift cards/certificates, toys, stuffed animals/Beanies, just to name a few. The silent auction is a means for the Foundation to raise a large amount of funds quickly. We are fortunate that at this conference, the local Foresters group will be helping us conduct the auction. Items may be brought with you to the hotel, or they can be shipped ahead of time. Sue Meckley (the grandmother of an individual with CHARGE) has kindly volunteered her home for the storage of items up to the conference. If you have questions, please see the detailed insert pieces.

We are still seeking "sponsors" for any of the meals during the conference. Meals and childcare are the largest portion of conference costs. Registration fees do not cover these aspects, and the shortage comes from the Foundation's general fund. Sponsors may support a full meal, or part of a meal. They will receive a public announcement regarding their sponsorship, as well as written acknowledgment in our program materials.

Should you have any questions, concerns or suggestions please contact Marilyn Ogan at 317-565-1419; 317-850-0712; marilyn@chargesyndrome.org; or oganm@insightbb.com.

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Tim is a psychologist who has done extensive research on CHARGE syndrome. He organized the Behavior symposium at the 2003 CHARGE Syndrome Conference in Cleveland. He has a 17 year old son with CHARGE.

A few years ago, my son Jacob, who has CHARGE syndrome, gradually stopped sleeping at night. The first night it took him two hours to get to sleep, the next it was three or four, and then he stopped. He would lie down, cover up, and then a couple of minutes later, he would be up out of bed. Our pediatrician could find nothing wrong, and suggested we get a psychiatric consult to find some medicine to deal with this new and troubling behavior. Knowing Jacob, my wife Nancy and I could not imagine that this was a psychiatric issue. We took him back to the doctor who upon further examination of the ear saw that what we had assumed was a piece of wax was actually a foreign object in his ear canal. We were unable to get an ENT (ours was on vacation) to do anything immediately, except to agree to schedule surgery. We finally took Jacob to the emergency room at the hospital where a physician (to whom I will always be grateful) pulled out the object (with three others of us holding Jacob down). We never found out what the object was, but it was impacted and infected and rubbing against his ear drum.

The articles you will read about pain in this issue and the next were initiated by the excellent article by Veronika Bernstein from Perkins School for the Blind. Veronika helps us to understand the categories of pain (acute and chronic), and how pain is processed by various systems (peripheral, spinal, emotional, and cognitive). She provides numerous suggestions for how to approach pain in our children with CHARGE.

I thought that a discussion of pain in CHARGE would be incomplete without a first person account. Chantelle McLaren eloquently emphasizes the critical role of parents in helping their children to manage pain with appropriate modeling, explaining, and supporting. It is through these significant relationships that children learn to manage their pain. Chantelle reminds us that pain can be both physical and emotional, and that both are extremely important.

David Brown highlights four important considerations regarding pain in our children. One is to understand potential involvement of the sensory system. He emphasizes the critical importance of recognizing that pain and medical issues need to be a top priority. He speculates about the role of self-regulation in the child's attempts to manage pain. Finally, David explores again the important role of parents in teaching the child to understand and cope with pain, as well in avoiding painful situations.

Lee Wachtel, a psychiatrist who is very interested in CHARGE, reminds us of how behavior is communication and that a failure to recognize what is being communicated can result in unfortunate consequences. So many children with CHARGE have communication impairments such that they cannot easily tell us "where it hurts."

In the next issue of *CHARGE Accounts*, Jude Nicholas, a neuropsychologist from Norway, will write about the environmental context to pain. If you have ever watched a toddler trip and fall, the child will usually look up at the parent. If the parent seems upset the child will burst into tears. If the parent smiles and says "oops," the child may smile as well. Our emotions are intimately associated with our interpretation of pain. Understanding the social context of pain can be important. Our body expresses itself through emotions, sensations such as pain, and behavior. Jude describes how educating children to understand pain can be an important component of pain management.

There are a number of common themes among these papers. Perhaps of most significance is a reminder to all of us of the critical role pain plays in the lives of our children with CHARGE. Knowing how to read our child's behavior, teaching our child about pain, preparing our child for potentially painful experiences, and providing constant support and encouragement are all important in helping our children to grow and to prosper.

Editors Note: *After you have read these articles, think about your child and his or her experiences with pain or discomfort. How did you know your child was uncomfortable? How did you figure out what was wrong? What strategies have helped your child experience less pain or more effectively communicate pain? Send your stories, thoughts and comments to Tim. He will collate many of them to go with Jude's article in the next issue of CHARGE Accounts.*

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CHARGE CAN BE A PAIN

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Veronika is with the DeafBlind program at Perkins School for the Blind, where more than a dozen students with CHARGE syndrome are enrolled and many more are evaluated every year.

We all have first hand experiences with brief episodes of acute pain from cuts and scrapes while growing up. However, pediatric studies report that chronic pain affects 15-20% of typically developing children. Kids can have painful ear infections that just don't go away, migraines, chronic digestion problems, and so on. Children with CHARGE syndrome are at a significantly increased risk of experiencing both acute and chronic pain. Development of the midline structures in formative stages (28-40 days after conception) is affected in CHARGE, along with the development of the middle part of the brain, which is involved with processing of pain. Multiple medical procedures may cause a habituation and decrease in pain perception as well as an increase in pain sensitivity. Medications may mask or disruptive behaviors may be a sign of pain. Generally speaking, pain is a multidimensional experience that includes sensations, emotions, thoughts, and feelings.

There are four mechanisms or levels of pain processing:

1. Peripheral: skin, muscle fibers
2. Spinal column: transmission of pain signals to the brain
3. Thalamus: emotional relay station of the brain
4. Cortex: multiple areas and sites of cognitive processing

We can experience pain on some or on all of these levels. For example, I may discover a bloody skin scrape after walking in the woods, and I may have no idea when and how it came about. This is a peripheral painful event that did not get to my emotional or to my thinking level. Pain processing systems change and develop throughout the life span. Individuals with CHARGE, in addition to being born with atypical systems, have many life events that change can their pain processing.

Studies of animals show that neonatal pain leads to structural and functional reorganization of the pain system. In general, early pain experiences decrease pain perception peripherally but increase brain-generated responses. A young animal may get desensitized at the skin level while stress and anxiety related behaviors are simultaneously increasing.

Many children with CHARGE show a similar pattern of decreased pain response to peripheral injuries on skin and muscle along with elevated levels of anxiety and anxiety spectrum behaviors.

Pain modulation also takes place on all four levels of pain processing. Potentially, we can modify pain processing on all four levels. Let's take acute pain. If bitten by a mosquito, I rub the spot. It helps to modify my pain perception. Lamaze method helps women in childbirth by teaching women how to process pain more effectively on all four levels. Lamaze trains women to rub the base of the spine (peripheral and spinal levels), to breathe (thalamic level), and to think imagine (cognitive level).

So, What Do We Do?

Acute Pain. I would suggest having a plan to address acute pain on all four levels by using a desensitization approach. This has been helpful for many of the kids with CHARGE with whom I have worked.

When there is a known scheduled potentially painful event, say a medical visit, use the child's cognitive skills to the max. Engage the child's thinking by telling a story in words, signs, pictures, and objects to prepare for the medical visit. Engage the child's movement and motor systems. If possible, do some "dry runs" to the medical office. Engage the emotional system. Plan for a positive mood-setting event prior to the visit by having positive objects and activities during the visit, and have a nice reward ready for after the visit. See the Summer 2006 issue of the CHARGE Accounts [ed. note: you can download this at www.chargesyndrome.org], for an excellent detailed description of a dental visit.

When there is an unplanned painful event (a fall, a scrape), do what your mother did. *Peripheral*: rub the spot, and apply cold or hot packs. *Movement*: pick up a young child, hug an older child, rock back and forth, engage in a familiar movement sequence (walk away, run, make up a game, do an errand). *Emotional*: kiss the pain away, model taking a deep breath; provide for emotional support and reassurance the way only you know best.

Chronic Pain. Chronic Pain is rarely peripheral. Most of the processing of chronic pain takes place at the spinal, emotional and cognitive levels. Adults are well placed to assess the possibility of chronic pain in most children. Pain detection is straightforward when your child is a good communicator, and you are sensitive to changes in your child's behavior and demeanor, even when those changes are subtle. If your child does not communicate effectively, a more formal assessment is

called for. Dr. Lynn Breau in Canada developed The Non-Communicating Children's Pain Checklist to be completed by caregivers. Unfortunately, some studies find that caregivers frequently underestimate or under treat pain. Please keep this in mind.

Pain medication is another complex issue. Ideally you would have a physician who is experienced not only with pain treatment but with the treatment of children with CHARGE syndrome. Daily life of kids with CHARGE often requires multiple and potentially conflicting medications. Also, there is no single medication that is appropriate for all levels of pain processing. Peripheral pain may respond to a number of medications. Chronic pain is chronic because it does not respond well to treatments. Chronic pain requires multiple trials of medications. We all are familiar with pain medications that reduce cognition, affect emotional functioning, and reduce motor functioning.

Have you noticed that the medical mainstream is getting more and more behavioral in the treatment of chronic pain in the adult population? This makes me very hopeful that we will continue refining behavioral and cognitive strategies in treatment of pain in our CHARGE world too. Hope to see you at the 2007 Conference.

PAIN: AN INSIDER'S REPORT

Chantelle McLaren

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Chantelle is a young woman with CHARGE syndrome.

Pain is difficult to explain. Parents worry how much pain their children with CHARGE suffer through both physically and emotionally during their lives. I could go on saying not to worry about pain and that pain is just something that we individuals with CHARGE get used to, but that would not be the complete truth.

I feel pain should not be ignored, but feeling sorry for the children doesn't help either. Parents need to explain to their children, even if their child doesn't fully understand. Tell your child what is going to happen if it's an operation. If it's bullying at school, tell your child that they must always come to you. You are their guide in life. If you show them pain is something to be afraid of, they will be afraid of painful situations; but if you show them that you are brave, they will see you are brave and be brave about the pain.

I can't speak for all individuals with CHARGE, but I have a very high tolerance to physical pain. I am tactilely sensitive but I have sprained an ankle, broken a rib and have had numerous operations without the need for a lot of pain medication. I don't even need any anesthetic when my dentist does work on my teeth because it doesn't bother me.

Emotional pain is too difficult for me to really explain. I have had a rough life with family and school. All I can really recommend is that parents work together to make their child feel good about themselves. If, heaven forbid, you divorce or are already there, please try to be amicable. No matter how deaf, blind or deafblind we are, one constant with CHARGE is that we pick up on EVERYTHING and internalize everything. I'm not saying shield your child; just always talk to them about issues in the family or the world. Even the littlest things can bother us, but with care and guidance from parents, caregivers, teachers and peers, pain can be managed.

SOME THOUGHTS ABOUT PAIN and CHARGE SYNDROME

David Brown

Education Specialist, California Deafblind Services

Many of you already know David from his many articles for CHARGE Accounts, many presentations at CHARGE conferences and other events, or talking with him at one of these events. He has made careful observations of individuals with CHARGE for many, many years.

As with everything concerning CHARGE syndrome, the subject of 'pain' is complex, multi-layered, and not very well understood. It is good that Veronika has brought up the subject, since it is often talked about but rarely written about. Here are my rather disjointed thoughts as a teacher with many years experience observing people with CHARGE but with no formal training in neurology.

Perception of pain for people with CHARGE

We know that almost everybody with CHARGE Syndrome has multi-sensory difficulties. Some of these are due to specific malformations of the sensory receptors that gather sensory stimuli (such as a range of eye defects, a variety of ear defects, or missing semi-circular canals). Other difficulties are due to neurological damage that causes sensory 'messages' to fail to be

conveyed to the brain or to be conveyed to the brain slowly, and in partial or distorted forms. Another contributory cause is brain damage that affects specific areas of the brain associated with receiving and processing specific sensory 'messages;' even if the messages arrive at the brain they may be missed or misunderstood. So if we consider pain as a sensory message, essential for safety and survival, we can see that there might be many occasions when a person with CHARGE may not feel pain as quickly, or with the same intensity, as other people. The situation becomes more complicated when we consider sensory integration issues, which might sometimes mean that the person's sensory system registers sensory inputs too powerfully, and other times not at all. With this kind of very poorly modulated sensory perception, some sensory inputs generally considered perfectly okay might be perceived as painful, so there could be a confusingly mixed pattern of over reaction to things that are not normally considered painful, but then under reaction to things that are normally considered painful and quite damaging, all within the same individual.

Causes of pain for people with CHARGE

For some time we have been telling parents and teachers that any deterioration in the behavior or functioning of a person with CHARGE should immediately lead to a consideration of health/pain/medical issues as a potential source of the deterioration. There are so many medical things that can go wrong and cause pain for people with CHARGE. Even a very partial list is daunting to consider. Some of the 'most likely suspects' would be ear infections, sinus infections, abdominal migraine, regular migraine, dental problems (especially impacting teeth due to irregular alignment or premature severe decay due to deficient dental enamel), difficulties with the autonomic system resulting in poorly regulated body temperature, digestion issues (especially constipation and/or gas), reflux of food or drink, the impact of persistent kidney reflux, the development of gall stones or kidney stones. A more recent addition to the list is seizure disorder, which we are just beginning to consider as a 'late-onset' anomaly in CHARGE syndrome; this might be preceded or followed by pain (for example, I have been told of people who experience headache or sore eyes as the first warning of an imminent seizure).

Recognizing pain in people with CHARGE

We are familiar with the pattern of sudden and apparently inexplicable upset in people with CHARGE, and although the possibility of pain should be investigated there could also be many other reasons. We should also investigate changes in sensory status (such as sudden deterioration in vision or hearing), new sensory challenges due to changes in the environment, and sensory over-loading. Problems with self-regulation are also frequently seen, and bring resulting difficulties with maintaining helpful arousal levels (I believe this is very common in CHARGE). The labile emotional state often reported in people with CHARGE could well be a reflection of these self-regulation issues – in the sense that many people with CHARGE might lack the sensory perception abilities to realize that they are getting stressed, anxious, or angry or are in pain until things reach a threshold, beyond which they perceive it with full force and kind of go nuts. I think of this as a lack of subtlety or gradation in their sensory perception abilities - either they feel nothing or they feel very strong inputs, but often nothing in between. The very high pain thresholds that are often reported in people with CHARGE (as when children never cry when they bump, never react to injections, and maybe stimulate themselves with very hard pinching or scratching or squeezing) are most probably a reflection of this issue.

Then there is a need to focus on external environmental factors when investigating why behavior is causing concern – these would include unexplained or unprepared changes in the environment, or changes in familiar personnel, or changes in familiar routines and so on. And here we must also consider communication issues as a part of the problem. The severity and complexity of multi-sensory deficit experienced by many people with CHARGE causes them to seek and demand a high level of predictability and familiar stable organizational patterns from the people and environments around them. When this stability and predictability is challenged the resulting confusion, frustration and fear can produce very dramatic changes in behavior that might look very similar to a person experiencing intense pain. With each individual how do we know what is causing the concerning behavior? 'Knowing the person,' or working to develop such knowledge, is a top priority, alongside medical investigation, as is having a forum for discussing hunches and ideas with others to reach a 'best guess' consensus.

If pain perception is thought to be poor

Veronika has included some good ideas in her article for dealing with perception of pain and with chronic pain. If pain perception is thought to be poor it is important to follow certain basic guidelines. At the most basic level it is important to protect a child as much as possible from potentially harmful situations until they begin to acquire the experience and skill required to navigate life's hazards safely. People with CHARGE might need help learning what things are dangerous and to be avoided even if they do not immediately result in a feeling of pain or discomfort. Examples might include avoiding touching knife blades or the points of sharp objects (forks, needles), not touching potentially hot objects (flames of any kind, stoves, cooking pots, heaters, hot water in showers or baths) without great caution or help from another person. Veronika mentions the idea of desensitization for pain; it might also be necessary to do the opposite and help to sensitize some people to perception of pain for the reasons I have already mentioned. For example, I am an enthusiastic supporter of Occupational Therapists who are flexible and creative in the use of the Sensory Integration Therapy approach and techniques, and a part of this would include

this sensitization/desensitization duality (both parts of which might be needed at different times for different reasons with the same individual). As well as helping individuals perceive and recognize pain, there may be a need to work on how pain or discomfort is communicated to others. There may be a need to introduce vocabulary about pain and for parts of the body, along with work to improve body image. I was recently told about a boy with CHARGE whose language was limited, and who had difficulty indicating on his own body exactly where he was hurting; things had improved significantly when he learned to point at a large picture of a body to indicate where he was hurting, and to use a 1-to-5 scale to indicate the degree or severity of the pain.

CHARGE and the MANY FACETS OF PAIN

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Lee is a child psychiatrist specializing in problem behaviors and psychopathology in children with developmental disabilities, with a specific research interest in children with CHARGE syndrome.

The routine assessment of pain has become obligatory in recent years across medical settings. Hospital accreditation organizations require such for all patients, regardless of age, cognitive or communicative levels. Health professionals are frequently taught to consider pain as the “Fifth Vital Sign,” following the critical variables of heart and respiratory rates, blood pressure and temperature. Those who have recently visited a physician’s office (what CHARGE parent hasn’t?) may even have been asked to sign documentation that pain was queried and addressed.

While pain is undeniably important for any patient, the potential presence of pain and the need for accurate assessment is particularly significant in individuals with CHARGE syndrome, who often present with a long history of medical challenges. Diligence is required for assessing susceptible sites and sources of pain, including ears, sinuses, mouth, upper airway, tracheostomy and gastric tube sites, gastrointestinal, genitourinary, cardiac and musculoskeletal systems, as well as old surgical sites and adaptive equipment. A frequent CHARGE staple, the seemingly benign hearing aide, may be at fault if it doesn’t fit correctly – one young child used to shriek and throw hers clear across the room – seemingly an uncooperative behavior until assessment revealed redness and tenderness of her ear from an inadequate mold.

Indeed, the effective communication of pain by a child with CHARGE, or any developmental disability with multiple impairments, is often the first stumbling block. While the child with CHARGE with adequate signing or verbal skills might be able to relate the “what, where, when and how bad?” of his/her discomfort, for most the task is not so simple. The challenge may be even greater when the child has differences in the experience of bodily sensory input. Nonetheless, clinicians have developed specific scales for those who cannot localize their pain, to rate it on a standard scale of 1-5. Most parents are familiar with the “face” scales, where a child selects from a range of joyful to calm to tearful to miserable faces. A parent or health care provider may also assess the child and choose the “face” most reflective of the child’s current presentation. Additionally, the Non-Communicating Children’s Pain Checklist-Revised (NCCPC-R) is a rating scale for children with cognitive and communicative impairments that adds six domains to facial expression, including Vocal, Eating/Sleeping, Social/Personality, Activity, Body/Limbs and Physiological, to provide a composite rating of acute pain.

While the NCCPC-R may be more useful in research than acute settings, its attention to various elements of a child’s overall presentation is key. Herein lie the clues to “what is different today about my child?” when there is no simple “point and respond” answer from that child. Red flags may immediately rise in the parent of a child with CHARGE who notices changes in these domains. A child who previously slept through the night without incident suddenly begins to rise and scream a few hours after his last meal. Is this an attempt to access mom’s and dad’s bed, is it an exacerbation of reflux, or is that pesky granuloma around the g-tube site irritated by the bed sheets? The previously cheerful, mischievous and energetic child suddenly begins to spend hours sitting alone, pressing her fingers into her eyes and face. Is this a new self-stimulatory or repetitive behavior, or is it a sinus infection? The child who was previously making great strides in school is suddenly quiet and withdrawn. Is there a problem with the classroom or peers, is there an infection brewing, or are these symptoms of anxiety and/or depression?

While the term “pain” typically evokes thoughts of physical distress, we certainly should not dismiss the possibility of emotional pain in kids with CHARGE, since research does support increased risk for psychiatric symptoms in CHARGE as well.

In teasing apart a child’s presentation and assessing the role of pain, an additional factor alluded to above and always worthy of consideration is the behavioral. Many children with CHARGE do have behavioral disturbances, ranging from compulsive and hyperactive behaviors, to self-injury and aggression, and many of these are fueled by causes completely unrelated to pain, such as additional psychopathology and environmental/operant factors. However, if behavior can be interpreted as communication, a frequent premise in CHARGE, behavior may be telling us an awful lot about pain and discomfort.

CASE I: A 5 year old boy with CHARGE syndrome had a long history of behavioral disturbance, including severe self-injury to his face, body and genitalia, as well as aggression towards his parents and caregivers and disruptive behaviors towards his environment. He was successfully treated through an inpatient behavioral admission with the combined usage of a structured behavioral protocol as well as psychotropic medication for compulsive behaviors and ADHD. Approximately 6 months after admission, he suddenly developed a severe increase in both self-injury and aggression for no obvious reason. He was seen by his pediatrician and diagnosed with viral pneumonia. Four weeks and a completed antibiotic course later, his behaviors had returned to baseline.

CASE II: A 10 year old girl with CHARGE syndrome was reported by her mother to have demonstrated low rates of self-injurious behaviors throughout her life, yet these were usually manageable with redirection. However, at discrete points in her life, she had developed severe exacerbations of self-injury, largely self-scratching of her face, causing significant bleeding and tissue damage. During each of these episodes, the child had to be hospitalized in restraints in pediatric intensive care while a full physical evaluation was undertaken. The culprit for her most recent episode was an acute bleeding ulcer.

CASE III: A 20 year old man with Cornelia de Lange syndrome, severe mental retardation and absent communicative abilities had never had a problem behavior in his life. Suddenly he began to engage in head banging against his wheelchair and biting of his hands. Caregivers were mystified until purulent discharge began to seep from his nose, and he was diagnosed with an acute sinus infection. Unfortunately, by this time the young man was also found to have a completely detached retina (from the head banging) deemed inoperable.

All three of these individuals lacked the ability to adequately report their pain via standard forms of communication, and thus turned to behavioral expressions, sometimes with severe collateral damage. This certainly supports the notion that parents, caregivers and health professionals must aggressively search for sources of pain and illness in individuals with disability. However, it also encourages us to actively develop ways of teaching our children to functionally communicate pain, or even the more rudimentary idea that something is “wrong.” A colorful example comes to mind of a young teen with autism with self-injury who had never developed any verbal skills but nonetheless had learned to use a Dynamo voice-output device effectively: when adverse physical side effects developed during a medication trial, he repeatedly pushed the “doctor” button until his parents contacted the physician and discontinued his new medication. As we work on developing our children’s basic life, self-care and self-help skills, communication of pain, discomfort or the simply “wrong” or “amiss” should be high on our list.

In sum, the issue of pain is of undeniable significance for individuals with CHARGE syndrome. The medical features of CHARGE syndrome make pain more likely in multiple body sites, and the potential need for surgeries, medical and adaptive equipment only compound the risk for painful experiences. The accurate assessment of pain is crucial, and often requires comprehensive assessment of overall changes in functioning and presentation due to communicative limitations. Not only must parents become experts in the detection of their child’s pain, so must the child be encouraged to express his pain and advocate for relief in a functional fashion, no matter how basic. Such proactive measures can help to ward off the development of problem behaviors as a method of communicating pain, while allowing for pain to be addressed promptly and comprehensively.

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 Frank & Joan Overton MI
In honor of Mallory Pigage
 Ellen P Elliott NY
In honor of Michael Kruger
 John & Cheryl Kruger MI
In honor of Michael Schifano
 Joanne & Sal Schifano NJ
In honor of Patrick Gately
 Pete & Vicky Smith IN
In honor of Peyton & Tate Young
 Tim & Carolyn Young MO
In honor of Sam Partridge
 Mike & Michelle Partridge AL
In honor of Sean Parker
 Marc & Sandra Parker IL

In honor of Wendell Wood
 Jane & John Garnett MO
 Joann Wood MO
In honor of William & Sandy Muir
 Wm & Patricia Newhouse PA
 Colleen & Phil Feather PA
In honor of Win Edwards
 Anne W Winter FL
In memory of Amy Nicole Arthur
 Wallace & Emmadell Vernon TX

And
 Ronald Podoll KY
 Wm & Andree Stanford NC
 Jeffrey & Ellen Schroy PA
 Brian Bartkowiak MI
 Alan & Martha Wismer PA
 Jeffrey & Lucille Harris VA
 Community Foundation for SE Michigan
 MI - Wade & Aliza Edwards
 Walter & Vivian Zagrobski MA
 Anne T Sturm MD
 Kenneth & Karen Wojcik MN
 David & Marty England AZ
 David & Barbara Cash MI
 Steven & Karen Miller NY
 Michael & Susan Arens VA
 George & Phyllis Saul NY

Last year the Board announced plans to establish an office at the Helen Keller National Center in New York. Although we have the office space, we do not as yet have a staff member on board. Meanwhile, we have closed the office at 409 Vandiver in Columbia and moved it back to my home at 2004 Parkade Blvd. Please use this address to send your conference registrations, membership fees, orders for materials, and donations to me at 2004 Parkade Blvd, Columbia MO 65202-3121. The phone numbers have not changed.

We are pleased to report that Lisa Weir has consented to serve on the Board of Directors for the next two years. She is assuming a position that was created by a resignation.

We hope you find the articles on PAIN informative and helpful. There will be more on the subject in the next issue. Please remember to send Tim your stories and comments.

Marion

*** * * Meet Our New Board Member - Lisa Weir * * ***

Hello everyone!

I just wanted to take a minute to introduce myself to anyone reading the newsletter who doesn't know me already, and to say hello again to old friends.

My name is Lisa Weir and I'm from New Brunswick, Canada. When I gave birth to my daughter Kennedy a little over nine years ago, our whole family— my husband Graeme, my sons Ryan, and Kyle, and myself— went through the typical “whirlwind” of receiving the CHARGE diagnosis. Our very first “connection” back then was to the US CHARGE Syndrome Foundation. I'll never forget the first time I reached Marion on the other end of that phone line, someone who actually *knew* something about CHARGE, who *understood*, and who was going to send us some much sought-after information. The Foundation was a bright light of hope for us back then, and has always been an important part of our lives since then.

Since Kennedy was quite young, I have been interested in

promoting awareness of CHARGE syndrome, linking families, sharing information, and personally learning all I can. I've been involved with setting up the national CHARGE syndrome group in Canada, with informal gatherings of the families in the Maritime provinces, and with CHARGE-related research in Canada. A lot of families also may know me from the CHARGE listserv, which I have been moderating for almost nine years. My latest personal learning endeavor is the completion of two undergraduate degrees, after which I plan on doing my master's degree in special education. My special area of interest, the education of students with CHARGE, will be the topic of my thesis.

I am thrilled to say that I've recently joined the Board of Directors for the CHARGE Syndrome Foundation. I am looking forward to the opportunity I've been given to give something back to the foundation, a wonderful organization that does so much to support families, for CHARGE syndrome itself, and that has meant so very much to my family.

In the Winter 2006 Issue of Accounts, we asked for interested members to apply for a position on the Foundation's Board of Directors. We explained that there were six positions available for persons to serve on the Board. When the deadline for applications arrived, we had received six applications. Since the bylaws state that “The six (6) persons receiving the highest number of votes will be elected,” it was decided that we would not hold a formal election but would declare these six duly elected.

We have printed the applications below from the new Directors as a way of introducing them to you. They will serve four year terms (2007-2011.) The other members of the Board who will be continuing to serve their four year terms (2005-2009) are Neal Stanger, Bonnie Haggerty, Marilyn Ogan, and Brownie Shott.

CHARGE Syndrome Foundation, Inc.
New Members of the Board of Directors

Lacey A Friedman, California Family member of individual with CHARGE

Hours available per week: 4-8 hours

Committee interests: Biennial Conference, Membership, Public Relations

Qualifications, Goals and Interests:

I believe I have some unique abilities that could serve other parents of children with CHARGE Syndrome. I hold a Bachelors of Science in Environmental Health and a Masters of Science in Hospital Administration. I am a researcher at heart and currently an inspector by profession. I am incredibly resourceful and assertive, and have an excellent ability to see both the minutia and big picture in projects/circumstances. But those credentials are not all that qualify me to be on the CHARGE Board. What qualifies me, I believe, is the dichotomy of both my private and professional life before Lucas, versus, after Lucas (my 4 ½ year old CHARGER).

The CHARGE Accounts newsletter is intended for general information only. Medical or treatment information and/or opinions are not necessarily endorsed nor recommended by CHARGE Syndrome Foundation, Inc. or its officers. Readers are reminded that the best source of medical advice is always their physician.

Privately....

Before giving birth to my amazing little Lucas, I was happily married to my college sweetheart. Life was great – great jobs, amazing recreation, wonderful home, etc. We waited until we had done everything in life we desired before starting a family – in fact we waited 16 years before starting what would be our “perfect” family. When we found out during pregnancy that our son would have medical problems and later that he had CHARGE, the professionals were excellent at providing all the surgeries and therapies that Lucas needed to survive. However, during those difficult years no attention was paid to the stress of CHARGE Syndrome on our marriage - consequently after 19 years together, our marriage ended. Since then I have worked to assist other new CHARGE families to help ease the stress and uncertainty. I have also pushed for counseling services for families of children with disabilities and hope to continue this advocacy work.

Professionally...

After graduate school I worked in hospital administration thereby learning the ins and outs of a hospital from the administrator’s point of view. I wrote policies, trained nurses, doctors and other staff, and provided recommendations to the executive officers. After Lucas, however I REALLY came to learn the ins and outs of a hospital from “the other side”! Now that I understand both sides, I am quite adept at working with the system to obtain all that my child needs. While I still find the authorizations, insurance, hospital stays, case workers, therapists, etc, burdensome, only occasionally do I find the “system” overwhelming.

I feel that I am very fortunate in being able to run for a CHARGE Board position as none of my son’s medical conditions are currently life threatening. Many CHARGE parents would probably love to be able to serve, but are not in a position to do so because of their child’s precarious health. I can, and therefore would - like to give back to those parents who cannot.

Matthew Murray, Massachusetts Family member of individual with CHARGE

Hours available per week: 10 hours a week or as many as needed

Committee interests: Biennial Conference, Fund Raising, Membership

Qualifications, Goals and Interests:

I have been on the board for the last 4 years and would love to remain on the board for many years to come. My son Justin was diagnosed with CHARGE Syndrome, when Justin was 15 months old, in August of 1993. Having a diagnosis helped me to identify and begin to reign in the many different doctors we had been seeing. It is important to me to help make sure that information is available to keep parents informed.

I have worked at the last five International Conferences, having been on the committee for the Boston Conference and a board member after that. I work for the Marriott Hotel chain and have a large knowledge of the hotel and meeting planning industry. I have served on many committees as a current board member. I see my strength as being able to help organize and facilitate the physical needs at a conference site. I also believe that fundraising is very important to the Foundation. Soliciting donation from various businesses and organizations is a daunting task to which we all need to work on.

Pamela J. Ryan, Massachusetts Professional

Hours available per week: 5 hours or whatever may be necessary

Committee interests: Biennial Conference, Membership

Qualifications, Goals and Interests:

I am the School Psychologist in the Deafblind Program at the Perkins School for the Blind in Watertown, MA. I have worked at Perkins for over 30 years in a variety of jobs all related to the education of children with deafblindness. As our population changed from a high number of students with deafblindness as a result of congenital rubella syndrome to a high number of students with CHARGE syndrome, my interest in CHARGE syndrome led me to the CHARGE foundation—becoming a member, attending conferences, joining the CHARGE listserv as an active member, and sharing information about CHARGE with our staff and parents. Additionally, because of the nature of my job here at Perkins, I have the privilege of meeting and corresponding with many families from around the country regarding their children as well as being part of their child’s evaluation should they come to our campus.

Being part of the CHARGE Foundation Board would enable me to further show my support for this wonderful organization in addition to being another voice—for my school and my own professional self—in the sharing of knowledge and learning regarding these very complex children and their families. The Foundation plays a crucial role in many families lives. I would like to work with the Foundation to further outreach efforts to families with older individuals with CHARGE and families of individuals with CHARGE who have multiple disabilities including significant intellectual impairment. This would give the Foundation another opportunity to create an even more inclusive organization.

My interests outside of work include a newly acquired love of tap dancing, beaches, walking, and spending time with the children in my life.

Lori A. Swanson, Tennessee Professional

Hours available per week: 5 hrs./wk. for work as a Board Member in addition to the 10 hrs./wk. I am currently spending on CHARGE research

Committee interests: Biennial Conference, Public Relations, Professional Services, Research

Qualifications, Goals and Interests:

I am a speech-language pathologist who teaches at the University of Tennessee (UT). In 2005, I attended the Conference in Miami Beach and decided that I wanted to make a major commitment to research on speech and language development in CHARGE. As a result, we have established a CHARGE Research Group at UT composed of Nancy Steele (NCDB), Jim Thelin, Wendy Keedy (from afar), and graduate students. I lead this group which is doing research on language development, hearing, and vestibular function in CHARGE. We will be presenting our work at the 2007 Conference. I also plan to present my research at the American Speech-Language-Hearing Association Convention in November in Boston. In addition, I am preparing a chapter on communication development for the CHARGE manual. At present, I am spending about 10 hours/week on CHARGE research.

At this time, I know that the CHARGE Board has only one member who is a professional. I am seeking a 4-year term on the Board to become even more involved with CHARGE and to make contributions as a professional. I enjoy the interaction with parents of children with CHARGE and with professionals who are interested in CHARGE. At present I am serving on the Conference Program Advisory Committee and I have begun as a member of the Research Committee. It would be an honor to serve the Foundation as a Board Member.

James W. Thelin, Tennessee Professional

Hours available per week: At present 20 hours a week

Committee interests: Biennial Conference, Professional Services, Research

Qualifications, Goals and Interests:

I am an audiologist who teaches at the University of Tennessee. I have been involved with the Foundation since its inception. Currently, I am the vice president of the Foundation and the only professional on the Board. My activities have included the following:

- (1) working closely with the President – especially on the relocation of our Central Office,
- (2) chairing the Professional Board and serving on the Research Committee,
- (3) participating in Conference planning and execution,
- (4) representing the Foundation to the National Consortium on Deaf-Blindness,
- (4) publishing research on CHARGE and writing service articles on CHARGE in publications for speech-language pathologists and audiologists, and
- (5) collaborating with faculty and doctoral students at the University of Tennessee in research on hearing, vestibular function, and language development.

I am seeking a second 4-year term on the Board so that I may continue the work I am doing.

John P. Wynne, Ohio Family member of individual with CHARGE

Hours available per week: 10 hours

Committee interests: I am willing to be on any committee that needs me

Qualifications, Goals and Interests:

I am a current member of the Board and have been for the past 3-1/2 years. I have been the Treasurer for the past two years. I would like to see the Foundation continue to grow in a positive manner to better serve its members. I would like to see fundraising continue to develop and one day see a national type fundraising campaign as well known as those for other disabilities.

Fundraiser in a Box is Fun and Easy!

Brownie Shott

Ok, so you've been wanting to do something for the CHARGE Syndrome Foundation but just weren't sure what you could do. Fundraiser in a Box is the answer! I held my Fundraiser in a Box pizza party event this past November and wanted to share how much fun we had and how easy it was to accomplish. Everything I needed came in the box from instructions to invitations to a pizza solicitation letter to thank you notes to the fabulous DVD which makes it so easy to communicate to your guests what CHARGE Syndrome and the Foundation are all about.

I approached a local pizza parlor who donated 4 extra large pizzas. If I hadn't gotten those, I would have gone to Little Caesar's Pizza where you can get large pizzas for \$5 each. Then, I bought a few large bottles of soft drinks, plates, cups and napkins and we were ready to go. I saved some time and postage by using Evite (<http://www.evite.com/>) for most of my

FUNDRAISING ACCOUNT

invitations. I scanned the invitation cover and used it in my Evite. I also sent some of the hard copy invitations to people who don't use email. We had a huge response and had more than 30 people show up.

I talked for about 2 minutes about how much the Foundation has meant in our lives, introduced Taylor (our son with CHARGE) to the group, showed the DVD and that was it. From start to finish was about an hour and people loved it. They loved learning more about CHARGE Syndrome and being a part of what we are doing. We raised over \$1500 and there is probably more to come.

Wouldn't it be amazing if 100 families each raised \$1000 for the Foundation using the Fundraiser in a Box! \$100,000 would make a world of difference in what the Foundation is able to do for families affected by CHARGE. Fundraiser in a Box makes it soooooo easy. You don't have to do a pizza party. You can do something more elegant or even more casual. The most important thing is you have the tools to have what ever kind of gathering you want, and the DVD tells the story for you. So, call the foundation office and order your Fundraiser in a Box. Have fun and help the foundation raise the funds it needs to grow and provide more support to families like yours and mine.

Brownie Shott

Katy, Texas brownie@chargesyndrome.org

Thank You to Everyone for Your Generous Support of the CHARGE Syndrome Foundation

Especially to

Altec/Styslinger Foundation AL
Ft Worth Volksfolks TX

General Fund

Austin Counter Tops TX
Jacqueline Durso CA
QuadGraphics & Jeana Phillips
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PA
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Barbara Johnson KS
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Employees Community Fund of
the Boeing Co WA

In honor of Aidan Gaucher

Mary L. Kramer MO
Kimberly Ruschmeier CT
In honor of Caitlyn McMullen
Cheryl & Joe McLaughlin PA

In honor of Charlotte

Cosenza
Mario & Shoshana Craig NY
Lindy Eye Care Staff NY
In honor of Clare O'Toole
Ronald & Laverne Krause OH
Rocky River Junior Women's
Club OH

In honor of Daniel Sheehan Ogren

The McShane, Ogren & Lukas
Families

In honor of Danny Schwartz

Estelle M Reich CA

In honor of David Ackerman Seicik

Allyson & Greg McGee TX

In honor of Erika Feather

Mark & Rajasi Mills PA

In honor of Ethan Wolfe

Joel Z Wolfe IL

In honor of Gracelyn Swann

Mary Louise Tesio CA

In honor of Jeanne Papesh's 55th Birthday

Susan Quigg PA

In honor of Makenna Susil

School fundraiser OH

In honor of Makenzie

Dougherty

Betty McKnight OH

In honor of Joel Saruski

Araly Herrera-Borgen FL

In honor of Megan Stanger

Richard & Lucille Goldsmith
NY

In honor of Michael Kruger

Ruth C Downey MN

In honor of Patty Haggerty

Nathan & Ryan Corcoran CT
Patricia A Bousquet CT

In honor of Reverend Glen

Pearcy

Anne T Sturm MD

In honor of Tiffany Grejda

Thomas & JoAnn Grejda PA

In honor of Zachary

Bluestone

Lois & Edwin Markowitz NY

In memory of Cedie Lynn

Mock

Kelly Kramer & Eugene

Marshall TN

Sarah & Jeremy Storrs WA

Eugene Marshall TN

Frances & Keith Ross NE

Carolyn Shelton TN

In memory of Corinne Ursula

Bernat

John, Cora, & Brooke Bernat

CA

In memory of Glenn Horne

Robert W. Shaw, Esq NY

In memory of Hope Peavey

Randy S Lewis GA

Faith Whitaker GA

In memory of Jacob Logan

Davis

Nathan J Milliron TX

Lisa J Steen TX

Kerry & Sharon Kirkland TX

In memory of Meghan

Graham

Julie & Don Graham MD

In memory of Michael Trapani

Laurence Beckerle NY

In memory of Ryota Yabuki

Munehisa, Akiko & Souma

Yabuki WA

In honor of Cody Johnson &

In memory of Wayne D Spear

Holly Lynn Johnson MI

In memory of Tyler Matthew

Bryony, Joe, & Hailey

SySantos DE

Fundraiser in a Box

John Belmont TX

Barbara Vissers TX

Kara Conrad TX

Jesse & Ginger Mosley TX

Harry Capers TX

Eugene & Kathie Gerritzen TX

Peggy Dicarali TX

Gary Minyard TX

Tom Shacklett TX

Derek & Julie Spier TX

Ardell Burns TX

Patricia Mead TX

Ron & Martha Aki TX

Lori Gunn TX

Kim Ginn TX

Terry Bieber TX

Katharine Temple TX

Conference

Chores-4-Charity

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Research

Scott & Anna Swanson VA

In memory of Meghan

Graham

Don & Julie Graham MD