



*Information and support for families of children with Hydranencephaly*

## Hydranencephaly Newsletter April 2008

This is our monthly newsletter about Hydranencephaly and the issues a child with Hydranencephaly might face. . Its purpose is to share information on the various aspects of Hydranencephaly as well as to show case our beautiful children. Much of the information presented in the newsletter will originate from the Hydranencephaly Mailing list or group which is hosted by Yahoo groups.

### Topic of the month:

Spasticity management part 1: medical treatments and surgery

### Spasticity Management

Spasticity or tone, is what will challenge you and your child. It can cause contractures, pull the hips out of joint and curve your child's back. But, there are ways to reduce the chance of these problems and ways to treat them should they occur. As this is such a huge issue for children with Hydranencephaly, I will dedicate 2 newsletters to it. In this edition I give information on medical treatments for spasticity such as Botox and Baclofen and on the surgeries many of the children need. Next month we'll discuss orthotics, positioning, therapy and equipment.

### Orthopedic Glossary

There are a lot of big words that you'll hear in reference to your child's physical development. Here are some of the most common ones.

**Abducted:** away from center of body

**Abductor muscle:** Any muscle used to pull a body part way from the midline of the body. For example, the abductor leg muscles serve to spread the legs away from the midline and way from one another.

**Acetabulum:** The area on the pelvis that is capable of becoming a hip socket. In newborns this area is cartilage, as the child matures this area will ossify or harden into bone.

**Adducted:** towards center of body

**Adductors:** tendons on inside of legs, they pull the legs together. Common surgery: adductor tenotomy cuts adductors to prevent legs scissoring.

**Adductor tenotomy:** The adductor muscle is used to bring the legs together. It is located across the groin. An adductor tenotomy is the term used to describe the snipping of the tendon that attaches to the muscle. This will allow the legs to fall further apart giving better movement of the child's leg so that the doctor can properly position the femur into the acetabulum.

**AFO (ankle-foot orthosis):** a brace used to stretch the Achilles tendon. Commonly made of a thin, light plastic material that is individually molded for a child. As a child grows, a new AFO may be needed.

**Asymmetrical:** One side of the body is different in attitude and action from the other. For instance,

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the head is always or frequently turned to one side, one arm remains flexed and fistled and the child does not use it. He may also kick with one leg.

**ATNR:** Asymmetric Tonic Neck Reflex: flexion in prone of side closest to the floor, extension of side away from floor. Generally it means that one side of the body is flexed and the other side is extended, therefore limiting the child's activities.

**Atrophy** Usually attributed to muscle, it is a shrinking in size, usually following a period of disuse or immobility

**Babinski reflex:** An important neurologic test based, believe it or not, upon what the big toe does when the sole of the foot is stimulated. If the big toe goes up, that may mean trouble. The Babinski reflex is obtained by stimulating the external portion (the outside) of the sole. The Babinski reflex is characterized by extension of the great toe and also by fanning of the other toes. Most newborn babies are not neurologically mature and therefore show a Babinski response. Upon stimulation of the sole, they extend the great toe. Many young infants do this, too, and it is perfectly normal. However, in time during infancy the Babinski response vanishes and, under normal circumstances, should never return. A Babinski response in an older child or adult is abnormal. It is a sign of a problem in the central nervous system (CNS), most likely in a part of the CNS called the pyramidal tract. Asymmetry of the Babinski response -- when it is present on one side but not the other -- is abnormal. It is a sign not merely of trouble but helps to lateralize that trouble (tell which side of the CNS is involved).

The Babinski reflex is known by a number of other names: the plantar response (because the sole is the plantar surface of the foot), the toe or big toe sign or phenomenon, the Babinski phenomenon or sign. (It is wrong to say that the Babinski sign is positive or negative; it is present or absent).

**Baclofen:** muscle relaxant used to treat spasticity. Can be administered as an oral dose or intrathecally, via pump. (Lioresal®)

**Botulinum toxin:** a drug, administered via intramuscular injection, to treat spasticity. (Botox®)

**Contractures:** permanent muscle and tendon shortening, a result of spastic tightening of muscles for long periods of time.

**Extension:** Stretching of any part of the body.

**Femoral osteotomy:** Cutting and repositioning the femur in order to hold the femur in the acetabulum.

**Flexion:** Bending of any part of the body.

**Hamstrings:** Muscles located in the posterior compartment of the thigh.

**Hip dislocation:** the hip is a joint where the upper

end of the thigh bone (femur) meets the pelvic bone. In children with spasticity, the end of the femur can gradually be pulled out of its socket where it connects with the hip. This condition is treated surgically

**Hypotonia:** (Floppiness): In some children brain damage leads to a state of tension of their muscles which is too low. They cannot therefore maintain any postures against gravity, and they cannot start a movement, maintain any postures against gravity, and they cannot start a movement.

**Kyphosis:** Exaggerated forward curvature of the spine, in the flexion/extension axis.

**Lordosis:** Curve of the spine backwards, whereby there is hollowing. Normal lordosis is seen in the lumbar spine, although variances do exist.

**Muscle tone:** The level of tension in the muscle.

**Osteotomy:** Osteo means bone, and otomy means the cutting of, therefore an osteotomy is the cutting of bone. There are two main classifications of osteotomies (pelvic and femoral), which are determined by the part of the body that is cut.

**Pelvic osteotomy:** Cutting and reshaping the pelvis in order to hold the femur in the acetabulum.

**Prone:** Lying face downward.

**Range of motion (ROM)** refers to the flexibility of joints such as elbows, wrists, ankles, knees, hips. Spastic muscles make joints stiff; this decreases the range of motion (span of movement possible) of the joint.

**Scoliosis:** Sideways (lateral) curvature of the spine (the backbone). The degree of scoliosis can range from mild to severe. Scoliosis is most commonly an incidental and harmless finding. Patients with milder curves may only need to visit their doctor for periodic observation. Persons with more severe scoliosis may require treatment. Severe scoliosis can often be improved by bracing, casting and/or surgical correction.

**Spasticity:** The increased stiffness or 'muscle tone' felt in children with cerebral palsy. Muscle spasm can often be felt when such a child's limbs are moved. The limbs are hard to bend or straighten because of the spasticity of the muscles. The doctor usually finds increased tendon jerks in a child with spasticity.

**Subluxation:** Partial dislocation of a joint. A complete dislocation is a luxation.

**Supine:** Lying on the back.

## Medical Treatment of Spasticity

Here is an overview of the most common medical treatments used to control spasticity.

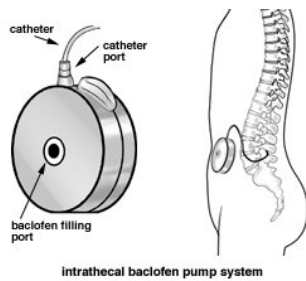
**Baclofen:** (Baclofen is also mentioned in the November 2007 newsletter but is presented in more detail in this edition.)

Information is from: <http://www.clevelandclinic.org/health/health-info/docs/0300/0369.asp?index=4590>

### Intrathecal Baclofen Pump System

#### What is baclofen?

Spasticity is a motor disorder characterized by tight or stiff muscles that may interfere with voluntary muscle movements. Baclofen is a muscle relaxant medication. It is commonly used in adults to decrease spasticity related to spinal cord injuries or multiple sclerosis. Baclofen is also used to treat children who have cerebral palsy.



#### How does baclofen work?

Spasticity is caused by an imbalance of excitatory and inhibitory input in the spinal cord. This imbalance causes hyperactive muscle stretch reflexes. These reflexes result in involuntary spasms and increased muscle tone. Baclofen works by blocking the release of excitatory neurotransmitters in the spinal cord. Ideally, baclofen restores the balance of excitatory and inhibitory input to reduce muscle hyperactivity. In this way, it also allows more normal motor movements.

#### Why is intrathecal baclofen preferred over oral baclofen?

Baclofen can be taken orally (by mouth in pill form) or delivered intrathecally (as an injection in the spine). Oral baclofen causes side effects that may limit its usefulness, especially when prescribed for children. Of the oral baclofen delivered throughout the body, only a small portion goes to the spinal fluid where it is needed to work. With an intrathecal delivery system, baclofen is delivered right to the target site in the spinal cord. Because the intrathecal system continuously delivers baclofen in small

doses directly into the spinal fluid, there are fewer and less severe side effects than that seen with the oral medication.

#### What is the baclofen pump system?

The baclofen pump system is the intrathecal (directly into the spinal fluid) method of delivering the medication. The system consists of a catheter (a small, flexible tube) and a pump. The pump--a round metal disc, about one inch thick and three inches in diameter--is surgically placed under the skin at the level of the abdomen.

The pump stores and releases prescribed amounts of medication through the catheter. The pump has a 10-mL fluid capacity and can be refilled by inserting a needle through the skin into a filling port (called the diaphragm) in the center of the pump.

#### Who is a candidate for the pump system?

Anyone who has spasticity and weighs over 30 pounds can be considered for the baclofen pump system. The pump system is appropriate when adjustable and potentially reversible treatment is desired, but cannot be achieved with oral medication.

#### How will my doctor know if the baclofen pump system will work?

If your doctor recommends the baclofen pump system, you will have a trial of the intraspinal therapy to test the potential effectiveness of the medication. During the medication trial, a small needle is placed near the spinal cord in the lower back. The medication is injected into the spinal canal and you are assessed over 8 to 10 hours to determine how well the medication treats the spasticity. If your muscles don't relax during the first trial, a larger dose may be given the next day to determine its effectiveness. This procedure causes minor discomfort. You may experience temporary muscle weakness due to the relaxing effect of the medication. This is caused by receiving a large dose of medication all at once during the trial. Patients who experience positive results with the intrathecal medication can decide with their doctor if they should have a baclofen pump system implanted.

#### How is the pump system implanted?

The pump is surgically placed just underneath the skin, usually in the lower abdominal area. The procedure to place the pump lasts from 1 ½ to 2 hours. The procedure may be performed under local anesthesia (pain-relieving medication that numbs the area while you remain awake) or general anesthesia (medication given by an anesthesiologist that induces sleep). Children are most often given general anesthesia and are asleep for the entire procedure.

The catheter is inserted through a needle and guided into the spinal canal, close to where pain pathways enter the spinal cord. The other end of the catheter is tunneled under the skin to the abdomen, where a pocket is created. There, the pump is implanted through an incision in the abdomen, anchored to surrounding tissue, and connected to the catheter. Once the incision is closed, the pump reservoir is filled with the dosage of baclofen prescribed by the physician.

#### **How is the medication dispensed?**

With a programmable pump, a tiny motor moves the medication from the pump reservoir through the catheter. Baclofen flows freely through the spinal fluid, affecting the nerves to control muscle hyperactivity.

Adjustments in the dose, rate, and timing of the medication can be made by your doctor, using an external programmer. The pump system can be set to dispense medication continuously or at certain times of the day. The pump can also be easily re-programmed if necessary.

Patients must return to their doctor's office for pump refills and medication adjustments, typically every 2 to 3 months. The pump system is taken out and replaced at the end of the battery's life span (which is usually 3 to 5 years).

#### **What are the side effects of baclofen?**

Some side effects of baclofen include dizziness, drowsiness, headache, nausea and weakness. Problems with the infusion pump can cause either overdose or sudden withdrawal of baclofen.

#### **What are the potential advantages of the baclofen pump system?**

Typically, the intrathecal baclofen pump:

- Effectively reduces spasticity and involuntary spasms, promoting a more active lifestyle, better sleep, and reduced need for oral medications.
- Continuously delivers baclofen in small doses directly to the intraspinal fluid, increasing the therapeutic benefits and causing fewer and less severe side effects than that seen with the oral medication.
- Can be individually adjusted to allow infusion rates that vary over a 24-hour period. It can be turned on or off or programmed to infuse different levels of medication throughout the day, depending on your needs. For example, people who find their spasticity helpful in maintaining leg extension for standing or walking can have a

lower infusion rate during the day.

- Can be turned off if spasticity reduction has shown no benefit.

### **Botox**

#### **Botulinum Toxin**

[http://www.wemove.org/spa/spa\\_bot.html](http://www.wemove.org/spa/spa_bot.html)

Two forms of BTX, type A (BTX-A, BOTOX® or Dysport®) and type B (BTX-B, Myobloc™ or NeuroBloc®) are commercially available. BTX temporarily weakens spastic muscles, allowing more normal limb positioning and function. To administer BTX, the physician places a small needle into the spastic muscle. Once the physician has confirmed the placement of the needle, the BTX is then injected.

The major side effects of treatment with BTX are dry mouth, blurred vision, excess weakness, or mild flu-like symptoms. BTX should be used with extreme caution in people with neuromuscular diseases such as myasthenia gravis or amyotrophic lateral sclerosis (also known as Lou Gehrig disease) or in those taking aminoglycoside antibiotics (including gentamicin, kanamycin, neomycin, streptomycin, and tobramycin).

One other important issue in the use of BTX in the treatment of spasticity is that of antibody formation. Because the immune system identifies BTX as a foreign substance, it produces antibodies that bind to and inactivate BTX, making it useless for spasticity reduction. Once a person forms antibodies to a particular type of BTX (A or B), any injections with that type of BTX are no longer effective. Repeated, high-dose injections are more likely to cause antibody formation than are less frequently repeated, low-dose injections. Therefore, the smallest amount of BTX necessary to achieve therapeutic benefit should be used, and the interval between treatment sessions should be as long as possible. Patients need to understand this important limitation on BTX therapy.

The effects of BTX are usually greatest for 2 to 6 weeks following treatment and usually fade completely after 3 to 6 months. In most situations, BTX is not injected again into the same muscle any sooner than 3 months after the last injection to decrease the possibility of antibody formation. In general, BTX cannot be used to treat

widespread severe spasticity, since the amount of drug required to bring about meaningful functional improvements would likely lead to antibody formation, resistance to BTX, and eventual loss of response.

There are less common treatments of spasticity which a few of the children have had. They are called Chemodenervation and Phenol and Alcohol blocks. For information on them go to:

#### **Chemodenervation**

[http://www.wemove.org/spa/spa\\_cd.html](http://www.wemove.org/spa/spa_cd.html)

#### **Phenol/Alcohol:**

[http://www.wemove.org/spa/spa\\_pa.html](http://www.wemove.org/spa/spa_pa.html)

### **Carly's Baclofen Trial**

Here's an account of Carly's (age 13) trial with Baclofen.

Carly's Baclofen trial went very well. She was put into a regular hospital room for the day, given an IV just in case she needed some emergency meds, and put monitor leads for respirations, heart rate and blood pressure. The hospital PT (whom we had never met) came in and measured her Range Of Motion and got a baseline on Carly's general spasticity. (Yes, she's very tight and has high tone, but is OK to diaper) A little later, they came in and talked with my daughter, making sure she understood the risks and what the procedure was all about. Then, they rubbed on the Emla (sp) Cream (to numb the area), put on a bandage and we had to wait for 45 minutes for it to deaden the area. The neurologist then did the spinal tap (lumbar puncture), checked her CSF pressure (which was good ... kids with high CSF pressure put stress on the pumps and they tend to have problems with them), and gave her a bolus of Baclofen. She didn't have a bad reaction to it at all. You couldn't tell she'd had any meds ...no breathing or bp problems at all. If she'd gotten too low on respirations or heart rate or anything, they were prepared to get her to the PICU, but she did fine. The Baclofen had a good affect on her leg muscles. She was relaxed and when the PT checked her the next 2 times, she had good results as far as tightness and ROM. It was amazing to see the PT lift one leg up high and the other leg stay on the bed. That's a strange sight. Carly's legs are always glued together :)

After the PT gave the neuro a thumbs up, we could go home. Now, we're waiting for the neurosurgeon to call with the surgery info.

I'm being very nice about the hours involved. Actually, we HAD to be there at 6AM without fail. They did nothing to her except the IV until 10AM ... ugh! ...the last part was over at 2PM. and then didn't come and report to Charlene after the procedure that she could take Carly home until about 5PM ... There wasn't any reason for holding us after 2PM, except that we think the doc forgot about us :( We're so glad we brought Carly's portable DVD player and lots of movies)

Overall, it was a great success. We were told it would be a 1 day thing, and it was. They never mentioned overnight unless she had problems. Maybe your doc does it differently.

### **What I've Learned**

The following are some points I've learned over the years about children who have problems with spasticity. It is taken from the book: Caring For Your Child With Hydranencephaly.

Unfortunately, no matter how much therapy and positioning we do with our children, they most likely will end up with some orthopedic problems. In this section I'm giving you some basic information on the more common ones and what can be done to prevent or treat them. Remember; I am not a physiotherapist or Dr, merely a parent. I am not promoting any particular treatment, just providing you information based on my experience and what is available on the internet.

#### **What I've learned:**

Before Kayda joined our family I had looked after many children and adults with severe disabilities. Many of these people had received very little or no physiotherapy or positioning and were very deformed and twisted. This becomes especially pronounced when the child goes through their puberty growth spurt. At that time, I vowed that if I ever had a child with a disability I would do everything within my power to prevent the deformities. It didn't matter how much time it took. Well, with Kayda I tried, and managed to prevent most of the severe deformities or problems, but not all. Along the way I learned a lot.

One of the things that you will be working against is a reflex called the Asymmetrical Tonic Neck Reflex. . The asymmetrical tonic neck reflex is a normal pattern of infant development, which is retained by many children with severe neurological problems. When the head is turned to one side, the arm and leg on that side (i.e., the face side) straighten or extend; the arm and leg on the opposite side (i.e., the skull side) bend or flex. The pattern is well described by its name, the asymmetrical tonic neck reflex. The pattern causes an unevenness of muscle action on the two sides of the body (i.e., asymmetrical). It causes an increase in muscle tone (i.e. tonic). The pattern is elicited by turning the head (i.e. stimulates receptors in the neck). It is a very automatic pattern over which the child has little control (i.e., a reflex). The ATNR leads to both structural and functional problems for children. Because the pattern is very strong in some children it can, over time, cause muscle contractures, hip dislocation or curvature of the spine; especially if the child's head is always turned to one side. It strongly affects children's ability to use their hands. Each time the child turns the head to look toward a hand or object placed on the side the arm on that side straightens and tightens. This makes it difficult to open the hand to pick up a toy and impossible to bend the arm to bring the toy to the mouth.

Something else that often happens to our children, and is in part related to the ATNR is that they get twisted over to one side. I.e.; with the limbs on one side in a flexed position they tend to pull or twist the child over to that side. If allowed to happen frequently this can have the child ending up in a "windswept" position-as they are twisted to that side it looks like the wind has blown them over to one side. I didn't know why this was bad until after Kayda's hips dislocated. In the windswept position both legs are pulled together or one may actually "scissor" over the other. This increases the likelihood of hip dislocation.

1. Do not let your child sit in a windswept position
2. Avoid "lounger" chairs that allow your child to sit twisted.
3. Get any splints or braces recommended for your child; afos (ankle foot orthotics), hand splints, abduction brace, back brace, whatever
4. Do "stretches" daily as much as you are able. These are range of motion exercises, which will be demonstrated by your child's

- physiotherapist.
5. Use a stander!!!! This is one of the most important things you can do to prevent your child's hips from dislocating. This chapter gives you the technical explanations for hip socket formation. I'll give you a layman's' description: When born the hip socket is fairly flat and shallow, it becomes cupped and holds the femur in place by bearing weight. Every minute in a stander helps to form a firm socket. It's not too late to start the standing after hip surgery either.
  6. If a dr tells you that your child needs to have the tendons at the top of her legs (adductors) snipped, do it. As long as the hip is still in place or only partly out (subluxed) surgery is called soft tissue and is relatively simple taking only about an hour and a half followed by 4-6 weeks in a cast. Once the hip is dislocated it becomes "bony" surgery (described in detail in this chapter) and is much harder on your child. My daughter had to have both hips reconstructed and it was awful. No one told me about preventing the hips from coming out by having the adductors snipped. Now I tell everyone.
  7. Watch your child's spine and how they are sitting. Make sure they are always positioned upright rather than hunched and try to make sure they don't twist to one side. Scoliosis & Kyphosis are real enemies for our children. Not only are they uncomfortable but both conditions can compress the lungs and compromise your child's breathing abilities.
  8. From what I've read; 95% of children with severe disabilities develop osteoporosis, which leaves them very vulnerable to fractures. When I heard this I was really concerned and wrote to a physio and a Physiatrist that I know. Their responses on preventing this were; prevent contractures (wear splints, position carefully), weight bearing (stander) and adequate calcium intake.
  9. The more your child uses a particular limb the tighter that limb will become-sort of a double-edged sword. With Kayda we could really see that as her hand use increased. There's not much you can do about it, it's just something to be aware of.
  10. Bones grow, muscles don't. For this reason, every time your child has a growth spurt their limbs/muscles will get tighter.

“Stretches”, standing and splints will help loosen them up again. I know that it’s hard to picture how these small steps can make a difference. With Kayda, at one point it appeared that she would need surgery to release her hamstring muscles. At the time she was still recovering from her 3rd hip surgery when she had nearly died so I was willing to do anything to prevent her from needing surgery. The physio mentioned previously suggested a soft splint by Medi Kids (address is at back of this book) worn at night and standing to lengthen the hamstrings. In only a few months she went from a contracture of –40 degrees to only –5 degrees. Just from those 2 strategies. She wore the soft splints each night and stood each day in her stander.

### Surgery

When all the preventative measures mentioned previously don’t work, surgery is necessary. Here is some information on the surgeries that our children often have.

#### Bilateral Adductor Tenotomy

This is usually the first surgery recommended for children with Hydranencephaly. Unfortunately I’ve not been able to find a good description of the surgery. It’s called a soft tissue surgery as it doesn’t involve the bones at all.

The tendons at the top of the legs are cut. This takes away the tightness in the legs. From what I’ve been told, the surgery usually takes about an hour. The legs are then put in a cast, either a hip spica which goes up over the abdomen and down to the ankles with the legs abducted (wide apart) or another type of cast or brace which keeps the legs spread apart.



Brennan had the above surgery as well as having his hamstrings lengthened. Brennan is in a Maple leaf brace and knee immobilizers.

#### Femoral Osteotomy:

A femoral osteotomy is performed when there is adequate coverage of the femur but still the femur can move with the possibility of dislocation. The

surgeon begins by opening the hip capsule. The femur is cut all the way across just slightly below the ball area. The surgeon then rotates the top of the femur slightly around towards the acetabulum. The femur is then put back together with a plate and screws. Lastly a hip spica cast is placed on the child, which remains on up to 8 weeks. The plate and screws are removed in a separate operation at a later date. The femoral osteotomy rotates the top of the femur around to fit better into the acetabulum (socket) while the bottom of the leg remains unchanged.

#### Pelvic Osteotomy

A pelvic osteotomy is used when the acetabular index is not improving and there is not adequate



coverage of the femur. The surgeon begins by opening the hip capsule up, hence many times the surgery is osteotomy with an open reduction.

After the hip capsule is opened, the surgeon will take a wedge shaped piece of bone from the bony protuberance further up on the pelvis. This is the graft bone. Next the surgeon will cut across the pelvis slightly above the acetabulum. The bone graft will be inserted into this cut and held in place with 2 four-inch long pins. Lastly the surgeon will place a spica cast on the child, which remains on up 8 weeks. The pins are taken out later, sometimes when the cast comes off and sometimes in a separate operation. The pelvic osteotomy brings the whole acetabulum (socket) down and around without changing the shape of the socket. There are different pelvic osteotomies depending on where the surgeon opens the hip socket (anterior, medial or posterior), where exactly on the pelvis the cut is made and whether more than one cut is made. But the resulting operation is still the same.

In the above picture Shane is in a cast after having an osteotomy done.

*Next month: Therapy, Positioning and Equipment*

Did I make the right decision?

All of us who watch our children go through orthopedic surgery wonder if the pain is worth it, or did we make the right decision. The following is a discussion on this topic from the Hydranencephaly mailing list which is hosted by Yahoo groups. It is used with the permission of the participants.

**Question:**

**Shandra, grandmother to Shane, age 3**

Maybe someone can help me with something I have been struggling with. The surgery was major, I did them when I was an OR nurse. Recovery is not fun. In Shane's case, it's not going to make him walk better, or even sit better. He didn't appear to be in constant pain from his hips, in fact he is usually quite happy. The only 'complaint' I heard was from the PT who didn't like his range of motion. Is this something that our kids have done because of what it will do if left untreated?? I just wonder if there wasn't something less invasive that we could have tried first.

**Answers**

**Dee Dee, mother to Elizabeth (1/17/89-**

**11/12/06)** Chandra, the reason ^ Elizabeth ^ had hip surgery was that she did show signs of pain when changing diapers or just trying to have her sit on my lap. That concerned us so we spoke to the Orthopedic doc and he said the surgery would ease her pain and get her hips back into place so that she would be able to sit comfortably. He knew this won't make her walk but he did know that she was very uncomfortable and he didn't like to see kids in that kind of pain. He also knew that ^ Elizabeth ^ just wanted to be able to sit assisted with a family member or her corner chair. He knew we would have done anything for her. You are doing the right thing for Sean. You make the decisions that you are comfortable with and don't worry about people. We are here for you.

**Ruth Anne, grandmother to Carly age, 13:**

I know you're second guessing your decision, but you shouldn't. Carly is facing some major work on her hips. Who knows, it may get all the way to hip surgery and casts. The way our doctor has explained it is that Carly won't be able to sit in her chair without pain for the rest of her life if we don't do something now. We're in line for the baclofen pump surgery at this point, but if her hip/hips come out of the socket before she gets

the pump she's going to have to get the pump and the baclofen going BEFORE the ortho will do the surgery on her hips. She'll just pull the hips out again with the high tone she's got in her legs.

So, we'll be making these hard decisions for our sweetie, too. They've never been easy. All you can do is weigh the benefits and the information you have and go for it the best you know how.

Barb, mom to Kayda (12/2/88-6/23/00): This is a dilemma that I faced with Kayda when she had her hip surgeries. Many people couldn't understand why she had them if it wasn't going to help her to walk. I had to explain that if we didn't fix her hips eventually she wouldn't be able to sit comfortably. With Kayda she did have pain from her dislocated hips. If the hips aren't properly aligned then they can make the twisting of the spine worse. So, she basically had her hip surgery to prevent future problems that would be harder to treat than the hips. I know it's a horrible surgery-Kayda had the full surgery twice and then had to have the first hip put back in place as well. If his hips were dislocated then the surgery he had was the only thing that would fix it. If the problem is caught before the hips are out then you can do things like the tenotomy of the muscles at the top of his legs or do botox or baclofen. I think that generally the "general" public thinks that walking is the goal for everyone and if a child isn't going to walk then why do surgery. Being comfortable and able to sit in a chair in the future is also a very good reason to have surgery. I'm not sorry Kayda had her hip surgeries. It made life much more comfortable for her and we weren't constantly dealing with the pain and difficulty of positioning her with her hips out of joint. It wasn't a pleasant surgery, nor was the recovery but how good things were in the long run made it worth it.

**Barbara, mom to Brianna, age 7:** Brianna has been in the hip spica cast 3 times now. Every time it was done I always felt like I had made the wrong decision. It was so hard to see her so miserable. After a couple weeks I started feeling better and when the cast came off and I saw the results I knew I had made the right decision. Just know that you made the right decision. Brianna now has perfect little hips that don't cause her any pain. During the whole process it was probably the hardest thing we have ever went through with Brianna. When I look at her now and know she can sit and stand comfortably; it really gives me peace of mind that I had made the right decision.

## Spasticity Care (in Heather Krueger)

Spasticity care is very important in children with special needs. It is important to keep the child's muscles loose so that they won't be in pain, so that they are easier to care for, and to help in their further development.

Children with hydranencephaly have notoriously mixed tone. They are "floppy" in the head and trunk, but very tight in the arms and legs. When Heather began **Early Intervention**, we were told about all of the problems she might have with her muscles tightening and were given a booklet explaining "**Range of Motion**" exercises that we were to do with her each day. Some of them she liked and some of them she definitely did not like. She worked very hard on them and would show off when, at the end of the week, the **Physical Therapist** would come back and do them with her. We would learn new ones each week and, in some cases, would get equipment to help us. While we were working on spasticity, we were also working on strengthening. Some of the items of equipment we had were a **stander**, a **therapy ball**, **wedges**, **DAFOs**, and **braces for her wrists**. We didn't get these all at once, but throughout her life we added them into her "collection."

As she got older, we were seeing that, while some areas were doing great (e.g., her feet, hands, and head), other areas were getting tighter. Her legs and arms were becoming increasingly tight. This made changing diapers, showering, dressing, and exercising a lot harder. We were afraid that we might end up breaking bones or dislocating a joint just to accomplish the daily routine. When it became increasingly clear that mere exercise wasn't going to do it, we notified the **Neurologist** and also started seeing an **Orthopedist**. This is when we got pharmacology on board. We started first with **Klonopin (Clonazepam)**. This wasn't much of a help. She became increasingly irritated and, of course, the irritability led to more spasticity. We weaned her from the Klonopin and started with **Baclofen**. Baclofen works for most of the kids but has the unfortunate side effect of lowering the seizure threshold. In some cases, the seizure increase is the lesser of the two evils and Baclofen is the way to go. Heather already was having quite a few seizures and we wanted to get them under control, so we also gave up on the Baclofen. Just as an aside, it is very important to wean slowly from

Baclofen. Stopping quickly can cause very serious problems.

We still needed help with her spasticity. Her Orthopedist had determined that her hips were subluxed, or partially dislocated. We were told they were out 33% and we would have to keep an eye on it. (We were also told that she had the beginnings of scoliosis. The Orthopedist said that a brace would not be a help for this, but we had a Boston Brace made from a cast to help her sit up straight and maintain head control anyway.) She was still allowed to be in her stander, but we noticed that she was crying quite a bit when she was in the wheelchair or going for rides. At this point we started going for **BOTOX** injections. BOTOX is another medication that can have bad side effects and might not have any beneficial effect at all. In Heather's case, the first round of injections worked slowly, but well. She was awake for the injections. Lidocaine cream was used to anesthetize the sites where the injections were given. Micro amounts were injected into the sites by the Orthopedic Surgeon, while Child Life distracted Heather with music and her favorite toy...the electric toothbrush.

As she got older and longer, the BOTOX lost its effectiveness and surgery was needed. Her hips were now both completely out, and she was in a lot of pain. We needed to resort to what was one of the most grueling surgeries of her life: **double osteotomies and tenotomies**. Her hips were put back into place and held with plates and screws. The tendons in her thighs were cut to keep them from pulling the hips out again. I often wonder what we might have done to prevent the need for this surgery, but it seems that we did what we could. I think that all of the running and climbing that a normally abled child does helps them, so possibly more constant and rigorous exercising might have worked, but we as a family weren't able to do that. At any rate, when the casts came off 5-1/2 weeks later, her muscles were very loose. It was hard to get used to holding her, as her legs were no longer like stiff boards but more like cooked spaghetti noodles. For the record, she cried a lot while the casts were on, and cleaning was a nightmare; however, she was no longer constipated. Her "poops" were as loose as her legs. She also cried a lot when the casts came off. She was on heavy-duty painkillers (such as OxyContin) for the first few weeks of this period. I think that it may have taken her as long as a year to recover from that surgery. We had her in the pool for **aquatic therapy** once a

week before the surgery and did it twice as often when the casts came off. This helped her to regain some muscle tone and didn't cause her much pain.

The high tone she had was gone and didn't come back, so the Orthopedist decided to remove the plates and screws. It looked like there was a formation around the left hip, and they thought that possibly the plate was rubbing and forming a bursa sac. (As a side note, if your child has plates and screws or any other metal hardware in his or her body, you should let the dentist know before you have your child's teeth cleaned so he or she can be put on antibiotics.) Within a month, she was x-rayed again and her left hip was coming out. The doctor was perplexed, but we decided that since she didn't seem to be in pain, we wouldn't do any more surgery.

In February of 2006 her spasticity was getting worse again. This time we decided to try **Intrathecal Baclofen Therapy**. For this, a tiny catheter is placed in the spinal fluid column, and a microdose of Baclofen is delivered directly into the spine. The catheter is hooked up to a pump the size of a hockey puck (and now even smaller) which holds batteries and a fluid chamber. The fluid chamber can be refilled by injection in the doctor's office. For many, this doesn't cause the seizures, because it is delivered in doses hundreds of times smaller than the oral dose and is delivered straight to the site needed instead of going through the brain. We signed up and had the trial done to make sure that the IBT would be appropriate for Heather. I was hoping for good results, but something went wrong with Heather's trial and her blood pressure dropped very low. She ended up in the PICU for three days until they were satisfied that her muscle tone and blood pressure were back to normal. She had gotten the smallest dose, and it was supposed to last for only eight hours. This was obviously not going to work for her either. Even if it had not affected her blood pressure, Heather's muscle tone would've been too low to allow her to be in the stander or to bear weight in any way without the risk of broken bones.

This is where the spasticity treatments ended for her. We continued to do the daily range of motions, but less than a year later she was discovered to have cancer. I asked to be allowed to put her in the stander. The Orthopedist came in and did some x-rays. They showed that her left hip was osteopenic—it was filled with cancer and

was now not dense enough to hold her weight. To put her in the stander would almost certainly result in a very painful broken hip. It was finally at this point that I realized that life as we knew it was over for Heather.

She had made some very great physical achievements for a child with Hydranencephaly, and I believe this was due to the hard work she did at home and in school with her physical and occupational therapists. With their help and the help of the doctors, we were able to, for the most part, control her spasticity.

Karen Krueger,  
Mom to ^Heather Joy Krueger^, (January 18, 2001 to March 2, 2007)

Heather on her scooter board, pushing herself



**April Birthdays**  
Luth: 4/8/96  
Logan: 4/9/00  
Ovrie: 4/10/04  
Zion: 4/18/07

**April "Sadaversaries"**  
(birthdays & anniversary dates for those who have died)  
Emma: **4/8/06**-12/1/06  
Kael: 6/6/04-**4/29/05**

Thank you to everyone who participated in this newsletter.