



Information and support for families of children with Hydranencephaly

Hydranencephaly Newsletter February 2008

This is the fourth edition of the Hydranencephaly Newsletter. It's 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.

Topics of the month:

Illness preparedness, Little Rooms, Children of the Month, Hydranencephaly Online

Things to consider if your child goes to the hospital

1. You are the expert on your child's care. You need to insist on being treated that way.
2. A family member needs to stay with your child at all times if possible. Hospitals usually don't know all the little specifics that go into the daily care of a child with high medical needs. And, with so many people involved it's easy for mistakes to be made.
3. Ask questions. Make sure you know what medications are being given to your child, what tests are being done and when.
4. You have the right to say no to having student nurses, drs. etc. "practicing" techniques on your child.
5. You are your child's voice. Your child has the right to be treated as a child, rather than a diagnosis.

"Go" Bag

What to have ready in case your child goes to the hospital:

1. Up to date list of your child's medications, doses, times, etc. I found it helpful to have all info that might be needed in an emergency on the same sheet with the medications. Things like legal guardian, allergies, code status, phone numbers of drs. and other professionals involved in your child's care.
2. Either enough medications for several days for your child, or have them in one place so that you can just grab that container as you head out the door. You can't always count on hospitals having your child's exact medications.
3. Care Notebook or similar document that outlines your child's needs, routines, etc. Something with an explanation of Hydranencephaly can also be helpful.
4. Syringes, feeding tube supplies, formula, any other supplies specific for your child.
5. Diapers/briefs especially if your child doesn't wear infant size diapers.
6. Reading material or something to keep you occupied. Anything you would need for at least 1 night away from home.

Table of Contents

Hospital Preparedness	1
"Little Rooms"	2-4
Make Your Own Little Room	5
Children of the Month	6
Birthdays and "Sadaversaries"	6
Consciousness in the Raw	7-8

Information about the Care Notebook:
<http://www.cshcn.org/resources/carentbk.cfm>

Stimulating your child “Little Rooms”

Now that Christmas is over, many of the people on the Hydranencephaly mailing list are thinking of how to stimulate their children more. There are also great sales on various lights and another nicely stimulating materials. One of the tools used by many of the families is something called a “Little Room”. It’s based on something created by Lilli Nielsen who is a Danish teacher-therapist. Her program is something called “Active Learning”. Children with Hydranencephaly can really benefit from this sort of stimulation. Of course an authentic “Little Room” is very expensive. Families have come up with their own versions which work just as well for their children.

In this section of the newsletter I’m including some basic information on the principals of active learning, some experiences and pictures of children using their “little rooms” as well as instructions on how to create this sort of learning environment for your child.

Active Learning is based on creating optimal environments for learners to actively learn on their own.

Design Principles of Active Learning

Equipment

The basic principles involved in designing the Active Learning environments are:

- **Feedback** - The environment must supply strong responses in multiple modes (auditory, tactilely, visually)
- **Support** - The child must be supported so as not to be in pain, typically not bearing weight on their legs fully or even partially
- **Richness**- Related to feedback, the environment must be thick with objects so that minor actions are rewarded with feedback
- **Variety** - However rich and responsive the environment is, it must be changed periodically-probably more often than we are inclined to think.

Above information is from:
http://www.lilliworks.com/new_page_1.htm

Experiences of families of Children with Hydranencephaly and “Little Rooms”

Noah, 6 1/2 Tasmania

When Noah got his first little room it was an actual little room that Lilli Neilson had designed - it had all the panels etc that you could interchange etc and was very expensive (early intervention bought it specifically for Noah to borrow). We then left early intervention and had to come up with something ourselves, and my brother is a builder. He just asked me to come up with the dimensions. I just thought about how long I hoped Noah would use it for (a VERY long time! :) and how big he would be, he made it accordingly .

Lisa and Noah

Noah’s current little room:



Here is another version of a Little Room which is used by Brandon.



From: <http://www.wonderbaby.org/articles/play-area.html>

What is a sensory play area?

The sensory play area is essentially a large box, often made out of PVC and peg board, in which small objects dangle from the lid so that the baby, when placed in the box on his back, can grab and play with the objects. The main concept is that this is a space where the baby feels comfortable and where he feels that *he is in control*. The baby can manipulate the toys and other objects and his voice will sound louder and different in the confined space.

The rules to the play area are simple:

- leave baby alone in the play area so he can learn about it himself
- keep the objects in the same place so that your baby will be comfortable and the space will be predictable
- use elastic to hang the objects so your baby can reach for an object, play with it, and then let it return to its position
- you can also attach textured fabrics, other materials, or even lights to the walls of your play area

What is it for?

The sensory play area is designed to...

- encourage reaching
- teach spatial awareness
- improve muscle tone in arms
- encourage vocalization
- teach object permanence
- teach babies that they can manipulate their environment, rather than wait to be manipulated by adults

What do I put in my play area?

Any household objects can work in a sensory

play area, from soup spoons to slinkies. Items in the play area should...

- be of pleasure to the baby
- be graspable
- have tactile and auditory qualities
- vary in weight and temperature
- take into account the senses of smell and taste
- be visually inspiring (if your baby has any sight)
- inspire to play counting games
- be changeable in shape
- be comparable

Here are some tips:

Think beyond simply hanging baby toys in your child's play area. When you do use toys, don't just use plastic ones; try hanging a metal slinky next to a plastic one so your baby can compare "alike but different" objects and learn to make his own comparisons. Also, blind babies are sensitive to the different temperatures of plastic vs. metal (warm vs. cold), and they find it interesting to compare the temperatures of both items!

Leave a little room between the hanging objects so your child doesn't become overwhelmed.

A small bottle brush that has been wet slightly is awesome - when your baby touches it or flicks it, a light spritz of water will gently mist him! Eventually, you can put the dangling bottle brush in a small 4oz. baby bottle so that he can learn how to put things in and take things out, but don't forget to keep the brush slightly wet so he gets the misting effect!

Krinkly gold metallic tissue paper is great fun! We bunch it up and tie elastic to the bunch.

If your child has light perception, string Christmas lights throughout the play area. Begin playing with the lights off. Then tell your baby that you're about to turn the lights on and watch their face light up as the twinkly lights fill up the room!

Instructions on how to make this sensory environment are on page 5.



Brennan and His Acti-Tunnel



Brennan got this for Christmas two years ago. It's called an Acti-Tunnel, it's similar to a Little Room in that it's like a private sensory room but "tent style". There are 12 rings to hang sensory-type items. I see no problem with hanging a string of lights around it. I like that it has a lot of room to accommodate a growing child as both ends are open, and it's wide enough to have a sibling or friend go in and join the fun. The big benefit is it's very lightweight and portable which makes it good for travel, or simple enough to just fold away for storage. Brennan enjoys his time in it, and we just think it's a great thing to have!



More info on the acti-tunnel

The tunnel is made up of light weight material. Half of it is solid and the other half is open mesh. Children can crawl inside on their own or be placed. Some children might like to be supine on a scooter board inside. Once inside in supine or sitting the child can explore the objects that hang from the 12 hooks that come with the unit. An optional set of 12 items can be purchased with the unit, or creative therapists, educators and parents can hang their own items. The unit folds easily and comes with a carry sack.

Sean's Little Room

Sean's room really isn't a built room.. I use an invisiboard, and a black blanket. It is in a half circle. He sits in it and i put light up things, as well as metallic, and sparkly things in there. All of them have Velcro on them. So they are removable.



For more pictures of the children using Therapy equipment see:

<http://www.hydranencephaly.com/therapyequipmentpics.htm>

How do I make a play area?

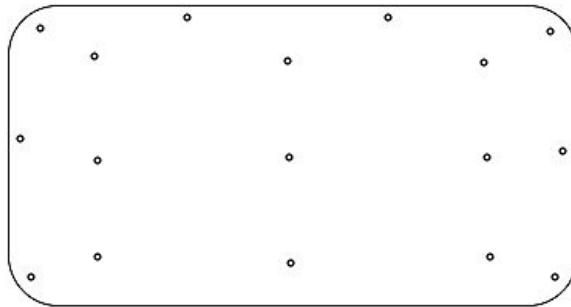
You can make your own sensory play area by following these simple directions...

Material's List:

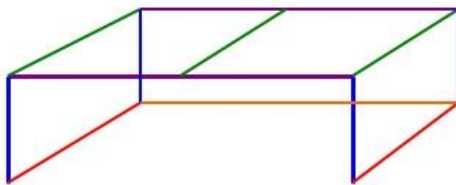
- 2 pieces peg board, 24"x17.5"
- 1 piece peg board, 36"x17.5"
- 1 piece plexi glass, 36"x24"—with 17 holes drilled through the top, following the diagram below (if you can't find or can't afford the plexi glass piece, just use another piece of peg board for the top)
- 3/4" PVC pipe, cut as follows:
 - 4 pieces 14" pipe
 - 2 pieces 20 3/4" pipe
 - 3 pieces 21 1/2" pipe
 - 4 pieces 16 1/3" pipe
 - 1 piece 34" pipe
- 2 90° PVC elbow socket fittings
- 2 tee PVC socket fittings
- 6 90° PVC side elbow fittings (SxSxS)—this piece is hard to find, you can also use...
- (option) 6 3/4 x 3/4 x 1/2 socket joints attached to 6 1/2 x 3/4 adapters
- cable pulls
- elastic
- household objects and toys

Instructions:

Take your material's list to the hardware store with you and have them cut all of the pipe to length. Have the plexi glass cut 36"x24" with rounded corners (for safety) and drilled as follows:



Using the color-coded diagram below as your guide, attach PVC pipes to make a box. (Here's a tip: Put a bit of sand in the legs of the PVC to weigh it down so it doesn't move or slide around the room.)



- 14" PVC pipe (4 pieces)
- 20 3/4" PVC pipe (2 pieces)
- 21 1/2" PVC pipe (3 pieces)
- 16 1/3" PVC pipe (4 pieces)
- 34" PVC pipe (1 piece)

Using cable pulls, attach peg board to back, right, and left sides and plexi glass to top.

Using elastic, attach your toys and objects. You're done!

Child of the Month

Bella



Bella is now 19 months old, we found out at 5 weeks she had hydranencephaly as her head grew rapidly. She had a shunt and ommya reservoir placed at 6 and 7 weeks respectively this has made the world of difference to our princess. Bella is full of character; she sees, hears and definitely knows what she wants and when, and how to get it. She has the most beautiful smile and it brightens everyone's day, she has fought many battles in her life so far and has proved her strength in every respect. We have physiotherapy, occupational therapy, speech and language therapy, hydrotherapy, osteopathy and now musical therapy which is her favourite. Bella has a standing frame, a corner seat and a wheelchair. Bella makes progress every day and we are so proud, she has learnt how to push the spoon away when she has had enough, and is getting really good at pulling her favourite monkey up to her mouth to kiss and suck. Bella has taught our family so much, most importantly to make the most of every day and show our love for each other. We are truly blessed. xxx

For more stories of
our children go to:

[http://www.hydranencephaly.com/
our_rays_of_sunshine.htm](http://www.hydranencephaly.com/our_rays_of_sunshine.htm)

February Birthdays

Braxton: 2/1/06
Nathan: 2/6/98
Charly: 2/7/01
Ada: 2/9/97
Brendan: 2/12/98
Lilly: 2/12/05
Carly: 2/13/95
Cobyn: 2/13/05
Troy: 2/15/05
Joshua: 2/16/06
Katie: 2/19/79
Marcy Ann: 2/19/00
Logan N.: 2/24/05

*Life is a
treasure given
to us all. It
doesn't have
to be perfect to
be valuable.*

From "Fighting for
David" by Leone Nunley

February "Sadaversaries"

(birthdays & anniversary dates
for those who have died)

Chris: 9/23/88-2/1/03
Noah M. 2/2/02-8/9/03
Jaime: 2/10/83-1/18/07
Chelsea: 11/9/00-2/17/01
Paiten: 7/22/04-2/19/06
Travis: 7/10/84-2/23/93
Lara: 2/23/06-12/10/06
Megan W.: 2/24/00-3/25/02
Blaine: 5/16/01-2/25/05
Amelia: 11/19/94-2/27/05
Julia: 2/28/00-9/25/00

"Angels" of the Month

Chris
9/23/88-2/1/03



Chris was always' my Valentine on Valentine's day. I could hardly wait to open his back pack to see what he made me. He always made neat Valentine's Day presents for me.

Chris had the most amazing energy. So full of life. You could see how happy he was, just by looking into those sparkling blue eyes. They gleamed with joy, most all the time. His smile and laughter, helped many people get through hard times in their lives, including mine. enjoyed everything we did. Including our visits to Florida to visit Grampy, every chance we got. They are my two best Valentine's. Now both in Heaven.

Madison

6/13/01-6/20/01

My daughter Madison...was alive just a few short days....seven to be exact and I think that the most I admired about her was the fact that she was a fighter...and she did some things that the doctors said she would never do. To me her biggest accomplishment was the day that she cried...they told me she would never cry and I wanted to hear her cry and I got to....it was a bitter sweet moment...I love you Madison love mommy.

What is being said online about
Hydranencephaly

Consciousness in the Raw

The brain stem may orchestrate the basics of awareness

Bruce Bower

In October 2004, Swedish neuroscientist Bjorn Merker packed up his video camera and joined five families for a 1-week get-together in Florida that featured several visits to the garden of childhood delights known as Disney World. For Merker, though, the trip wasn't a vacation. With the parents' permission, he came to observe and document the behavior of one child in each family who had been born missing roughly 80 percent of his or her brain.

These children, 1 to 5 years old at the time of their Disney adventure, had suffered strokes as fetuses or had experienced other medical problems shortly before or after birth that destroyed nearly all of the brain's outer layer, or cortex. In this rare condition, called hydranencephaly, cerebrospinal fluid fills the gaping hole within the child's head.

Such youngsters often die in the first year of life as a result of seizures, cerebral palsy, lung abnormalities, and a variety of other physical ailments. With proper medication and the installation of shunts to drain fluid from the braincase, however, some individuals live 20 years or more.

Neurologists typically regard hydranencephaly as an anatomical sentence to a lifelong "vegetative state." Such children supposedly validate a brutally simple equation: Little or no cortex equals no awareness of any kind. In family activities observed in the Magic Kingdom and elsewhere, the kids quickly cast doubt on that standard assumption. Merker noted that these cortex-deprived, nonverbal children remained alert for much of the day. They reacted to what happened around them and expressed a palette of emotions. A 3-year-old girl's mouth opened wide and her face glowed with a mix of joy and excitement when her parents placed her baby brother in her arms.

The youngsters displayed good hearing but limited eyesight, a curious pattern given that they typically retained small parts of the visual cortex but none of the auditory cortex.

In observations at each child's home, Merker noted that these youngsters recognized familiar adults, liked familiar settings, and preferred specific toys, tunes, or video programs. Although

saddled with limited mobility, some kids took behavioral initiatives, such as learning to activate a toy by throwing a switch.

In the February *Behavioral and Brain Sciences*, Merker, an independent neuroscientist in Segeltorp, Sweden, described how the accomplishments of these children relate to behaviors recorded in prior studies of human-brain function and of animals after surgical removal of the cortex. His analysis generates a provocative proposal: Basic awareness of one's internal and external world depends on the brain stem, the often-overlooked cylinder of tissue situated between the spinal cord and the cortex. Merker argues that the brain stem supports an elementary form of conscious thought in kids with hydranencephaly. It also contains auditory structures capable of preserving hearing in someone without a cortex. In contrast, optic nerve damage in hydranencephaly frequently impairs vision, regardless of what the brain stem does.

Self-awareness and other "higher" forms of thought may require cortical contributions. But Merker posits that "primary consciousness," which he regards as an ability to integrate sensations from the environment with one's immediate goals and feelings in order to guide behavior, springs from the brain stem. If he's right, virtually all vertebrates—which share a similar brain stem design—belong to the "primary consciousness" club. Moreover, medical definitions of brain death as a lack of cortical activity would face a serious challenge. At the very least, physicians could no longer assume that individuals with hydranencephaly don't need pain medication or anesthesia during invasive medical procedures.

"To be conscious is not necessarily to be self-conscious," Merker says. "The tacit consensus concerning the cerebral cortex as the 'organ of consciousness' ... may in fact be seriously in error."

Brain drain

(this section is very technical. To read it, go to: <http://www.sciencenews.org/articles/20070915/bob9.asp>)

Cortical divide

Of 27 comments by mind and brain researchers published with Merker's article, nearly half agreed that the inner workings of consciousness lie in the brain stem.

"The roots of consciousness exist in ancient neural territories we share with all vertebrates," says neuroscientist Jaak Panksepp of Washington State University in Pullman. "By the weight of empirical evidence, all mammals are sentient beings."

In his own research, Panksepp studies the ability of animals to experience biologically based states of

mind or feelings that range from hunger and thirst to emotional delight and distress. For instance, Panksepp and a coworker reported in a controversial 2003 paper that rats express "joy" while playing with other rats by making ultrasonic sounds that represent an ancestral form of laughter.

Psychologist Carroll Izard of the University of Delaware in Newark emphasizes that this form of primary consciousness, as Merker would put it, or "primary affect," as Panksepp terms the rats' consciousness, consists of sensory activity in the brain stem. This capacity generates emotions and an awareness of one's surroundings but not an ability to talk about what one has experienced, Izard continues. In the same way, people can become conscious of a feeling that they can't label or describe, a phenomenon that's especially common in healthy infants and in children lacking a cortex, Izard says.

The existence of primary consciousness challenges widespread assumptions among physicians that newborns and fetuses can't feel pain, adds pediatric neurologist K.J.S. Anand of the University of Arkansas for Medical Sciences in Little Rock. Evidence now suggests that adult and immature brains use different systems to process pain, Anand says.

The brain stem and the thalamus, a relay station for sensation just above the brain stem, foster pain responses in babies before and after birth, he asserts. The cortex takes over pain perception as it greatly expands during childhood and adolescence, Anand hypothesizes.

Other investigators criticize Merker for denying the cortex its traditional position as the brain's engine of consciousness. Even if a basic form of consciousness exists, they regard it as at least a partial product of the cortex, not just the brain stem as Merker argues.

Conscious thought probably relies on the workings of connected brain areas within and outside the cortex, contend Susanne Watkins and Geraint Rees, neuroscientists at University College London. "It seems unlikely that activity in any single area of the human brain will be sufficient for consciousness," they write.

Children with hydranencephaly studied by Merker possess remnants of cortical tissue that could have triggered states of awareness, the researchers suggest.

Other commenters, including philosopher Gualtiero Piccinini of the University of Missouri–St. Louis, cite prior evidence that the cortex by itself regulates visual awareness. Following visual-cortex damage, certain patients report no

conscious ability to see on one side of their visual field but still unconsciously perceive the identity and location of items in that same visual field. Scientists call this phenomenon blindsight.

The most extensively studied blindsight patient has frequently reported being aware of "something" in his blind visual field, Merker notes. This man retains primary visual consciousness of his surroundings but can't describe what he sees in words, the Swedish researcher contends.

Reclaimed kids (*this section relates part of the 1999 report by Dr Shewmon. To read this go to: <http://www.sciencenews.org/articles/20070915/bob9.asp> or see his article on the Hydranencephaly website*)

"If these children had been kept in institutions or treated at home as 'vegetables,' there can be little doubt that they would have turned out exactly as predicted," Shewmon says.

After making his own observations of children with hydranencephaly and their families, Merker seconds that point. He notes that well-treated youngsters born with little or no cortex regularly display brief losses of consciousness due to absence epilepsy, a clear sign that at other times they're conscious.

Parents described these lapses of awareness in their children to Merker with phrases such as "she is off talking with the angels."

Perhaps most intriguingly, kids with hydranencephaly demonstrate that the brain stem is not simply a reptilian relic stashed in the brain's basement. "The human brain stem is specifically human," Merker says. "These children smile and laugh in the specifically human manner, which is different from that of our closest relatives among the apes." For now, the neural puzzle of consciousness remains unsolved. But cortically endowed investigators may have much to learn from cortically deprived kids.

This entire article can be read at:
<http://www.sciencenews.org/articles/20070915/bob9.asp>

To read the preprint version of Dr Bjorn Merker's Article "**Consciousness Without a Cerebral Cortex**" go to:
http://www.bbsonline.org/Preprints/Merker-03062006/Referees/Merker-03062006_preprint.pdf