

Cognitive Development and Awareness in a Child with Hydranencephaly

According to a parental survey conducted from 2005-2007

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Hydranencephaly Information Network

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Purpose statement: Hydranencephaly is a rare neurological condition in which all or most of a child's brain dies and is replaced with fluid. Common assumption is that a child with Hydranencephaly is not only incapable of learning, but is unconscious and totally unable to interact with his/her surroundings or people in his life. In 1999, Dr Alan Shewmon, a neurologist in California published an article entitled "Consciousness in Congenitally Decorticate Children: "Developmental Vegetative State" as Self Fulfilling Prophecy". He gave examples of 4 children with Hydranencephaly and their development and awareness. His premise was that if a child was treated like he/she was in a vegetative state and given no stimulation, then he would appear to be in this state. In 2007 Brain and Behavioural Sciences published an article by Dr Bjorn Merker. It's title was "Consciousness Without a Cerebral Cortex: A challenge for Neuroscience and Medicine. Dr Merker used video and pictures of a number of children with Hydranencephaly to show that consciousness and development is possible in those who only have a brain stem. Both of these articles only cited a few children as examples. Since 1998 a steadily increasing number of families have been corresponding via email as part of a group named "Rays of Sunshine". A lot of information has been shared by these families. At this time there are over 300 individual children whose family we've been in contact with. As parents of children with Hydranencephaly we've shared many examples of our children doing things that wouldn't normally be expected in children who are missing all or most of their cerebral cortex. But, there have been no firm numbers to back up our claims. In order to get a clearer picture of how children with Hydranencephaly interact with and respond to people and their surroundings a survey was conducted via an online survey hosting site. This survey consisted of a series of multiple-choice questions about various signs of awareness, along with an open-ended question asking how the child shows this awareness. The survey was online from 2005-2007 and had 106 participants. This is the largest sample of children with Hydranencephaly and it is hoped that in compiling this data we'll be able to show the true capabilities of a child with Hydranencephaly.

The vast majority of children with Hydranencephaly are very alert and aware and in some ways much more alert and capable of learning than a child who is born with a complete brain that is then severely damaged. We realized over the years that although we feel our children are able to learn, they always do poorly on any assessments they're given. It should be noted however that those assessments are developed for mobile children with a full cortex.

The purpose of this document is to answer the questions:

- a. Is a child with Hydranencephaly aware of his/her surroundings? This includes awareness with regard to objects, people, and showing they know the difference between familiar people and strangers.
- b. Can a child with Hydranencephaly learn?

As a child with Hydranencephaly usually has limited motor skills and no formal speech, how would you know if a child is learning? As will be seen in the results of this survey, children are very good at indicating that they're learning and aware. What signals or indication can children such as these give to indicate this? I hope to show that learning can be indicated by a change or increase in reaction or interaction over time.

Kayda and her answers to the survey

Kayda was born in 1988 and died in 2000 at the age of 11 and a half. She had Hydranencephaly. She came to live with me at the age of 4 ½. When she was first placed in my care she was almost totally motionless. By the time of her death seven years later, she had a whole range of movements and expressions. These are my observations of how Kayda became that person.

When Kayda came to live with me at the age of 4 ½ she rarely moved her hands, and hardly moved her legs. She also struggled for every breath. Once I realized that her breathing was better lying down and I kept her in that position most of the time, Kayda started to change and move.

The first evidence of growth in this area was when she started moving her hands or arms enough to dislodge whatever stuffed toy was under her arm. At first I didn't believe she was having purposeful movements as I'd been told everything she did was a reflex only. One of the earliest deliberate things I remember her doing was running her hands over the side of the bathtub as if she was comparing textures. You're just imagining things, I told myself.

That Christmas I had a hard time finding toys for her, but I found a few that could be activated by very little movement (by accident) easily. One was a "Dinoraur" (Bruce) by Fisher Price. It made a noise when squeezed and I hoped that eventually Kayda would learn how to squeeze it purposefully. One day shortly after Christmas I watched her turn her mouth to "Bruce," turn away, and then turn back and start to chew on him. That simple act was the beginning of a whole new life for Kayda.

Within days she was stroking Bruce and anything near her, and chewing on anything she could get near her mouth. Soon she was actively touching toys and figuring out how to manipulate them. She stroked them, batted them, and pushed them onto the floor from her tray if she didn't want to play with them. From that day on, she always had her hands raised and searching for things. As time went on she became more aware and more selective as to what she held on to or played with.

Throughout her life, Kayda remained unable to move much, never spoke any words and had many health issues, but starting sometime after her 5th birthday which was at the beginning of December, she became increasingly aware of and interested in her environment.

She also showed an awareness of her own body. She stroked things, held her own hands and discovered her feeding tube, constantly touching and playing with it. It often looked like she was comparing textures and body parts. She'd touch one part and then touch another and then go back to the first. She did this with clothing and toys as well.

As she got older, Kayda sometimes seemed almost driven to touch things. At other times, especially after her surgeries, she was in too much pain to play with her hands. She stopped using her hands almost entirely for a year after her first hip surgery at age 6 ½. It wasn't until after the 2nd surgery at age 7 that she more or less got back to "normal." After that, I remember her being driven to touch things and her hands (like wringing her hands) just after her 3rd hip surgery at age 8 and some other times when she wasn't well.

Although she started to show that she knew the difference between familiar people and strangers within a month or so of coming to live with me (July), this really increased about her 5th birthday (December). She showed this by smiling more readily at those she knew well. As time went on she got much more responsive to me and when she wasn't well, she did not want me to leave her side. At her sickest, I had to hold her constantly. She'd stiffen up if I even moved in a way that made her think I was putting her down. She maintained this level of awareness as she got older. It remained consistent. The number of people she got to know and respond to did grow.

I could always tell when she recognized something or someone; her eyes would stop moving back and forth and it appeared she was listening. She started it at the age of 5. As she got older she clearly recognized more people and things. This included movies and stories as well as people. She showed the same type of recognition of familiar voices starting at about the same age. For instance, she loved the man who gave the sermon at church. She'd let out a little chirp of protest when he stopped speaking. One day when we went to a Memorial service for a family member, Kayda fussed and fussed during the message. I finally realized that it was the "wrong person" telling what she felt was a story. She expected the regular man. She maintained this level of awareness even while the number of people she responded to increased.

When needing comfort or reassurance, she would let only me comfort her. At one point, she went through a period where she was easily startled. This could go on for hours and would only end when I picked her up. One day I got called to the school as she'd gotten startled on the bus and could not be calmed. I went in and picked her up and she settled down right away. After a few minutes she lifted her head off of my shoulder and she went "ah" in a defiant tone as if to say "see, I told you my mom would make it better." She was then completely relaxed and I was able to leave her at school. There were also several times throughout her life when she was very sick and not expected to survive. If I picked her up, it had a restorative effect and by the time I put her down, she was on her way to recovering. She and I had a very strong bond that lasted until her death.

Kayda showed no indication of fear or dislike; however, she did show a strong aversion towards the use of her nebulizer mask. She would become very agitated if the mask was placed on her face. She would start to breathe more rapidly and have a frantic look on her face. This started at the age of 4. The only time she would tolerate the mask is when she was very ill. I knew when she was getting better because she'd start to fight the mask again.

She also showed a strong response towards certain stories. She started to listen to stories at the age of 6. Her favorite stories were a series called "Classical Kids." They were stories about the composers with their music. She'd always fuss if you put on a story about Tchaikovsky or Bach. She just liked the Mozart, Beethoven and Vivaldi stories. The stories about Bach & Tchaikovsky were just the "composers" talking to a child. The Mozart, Beethoven, Vivaldi & Handel stories were actual stories with a plot involving children and the music. She seemed to know the difference. As she got older, her story preferences changed. She would make it very clear that she didn't want to listen to a particular story by fussing and not settling to listen to the story until I'd put a different one on.

Although she would fuss to make her wishes known, she only had a temper tantrum once. It was after school. As we had to go out later in the day I wanted her to lie down on the couch for a rest. She objected strongly and started crying. She cried and cried, and of course didn't sleep at all before we went out. She eventually stopped crying.

Her preferred toys were anything that she could touch or manipulate including "tactile" books. These were books made by the children at school. They had a story and a picture with it. The picture was made of various seeds and foods glued to the page. She felt it was her mission to get everything off of the page and on to the floor. She also liked a variety of toys, most of which were sold as "baby" toys. As she got older she started to refuse to play with anything that she felt was for a younger child or too simple for her. She liked the toys that had several different ways to activate them. She seemed to know the difference on her own. She'd push those rejected items onto the floor right away. She eventually only wanted to play with a keyboard and a toy with the alphabet on it. They used the same sort of hand actions, but she knew they were more appropriate for her. I don't know how she knew; she was just very consistent on what she would play with and what she rejected. She started this selectiveness at about the age of 9.

Kayda's favorite activity was to watch videos (when her vision was tested at the age of 7 or 8 it was found that she could see a lighted object about 5 feet from her) or listen to books on tape. Her love for stories started just after her first hip surgery at the age of 6 ½. I'd been afraid she'd lose skills or awareness after the surgery due to the pain. She did stop using her hands much for about a year, likely from pain. One day while she was still in the hospital I put on a tape that had some stories on it. I could tell she was listening to it as the movement of her eyes changed. I played some other stories I had borrowed and her reaction was the same. By the time we left the hospital she'd fallen in love with listening to stories. Previously she'd shown no awareness of or interest in stories. Her interest in videos was also limited prior to that. This appreciation continued to grow and develop up until just before her death. As she got older she changed from liking the younger type videos and stories to liking adult ones. She'd fuss if I put on a short story written for early readers and would settle once it was an adult based book. She loved the Dragon Riders stories by Anne McCaffrey. She also shifted from wanting to watch Disney movies to only wanting "adult" ones like Star Wars and Star Trek. The Indian in the Cupboard series were her favorite books and the movie her favourite film. She had all 4 of the books read by the author. Another change I saw in her as she got older was that she seemed to know when a book had more than one tape to it. If I put on a multi tape book at bedtime she wouldn't go to sleep until she'd heard the entire book. I know it doesn't seem possible, but I and many others tested her on this a number of times and every time she stayed awake until the end of the book. It seemed like she was aware of the story and if it had ended or if it was just stopped.

Her favorite pieces of equipment were her stander (a piece of equipment with straps that allowed her to stand), her wheelchair and her comfy chair (a lounge type chair made just for her). She would settle down if she was in one of these pieces of equipment and fuss if she wasn't. When she was younger she hated to sit, she was only happy lying down. As she got older she decided that lying down wasn't for her except at night time or for her nap. Towards the end of her life she was only happy in her comfy chair (a

custom built lounge chair). At that time, she was only comfortable sleeping and that's what she did whenever she was in her comfy chair.

Kayda had a number of security items. Her first two were the "dinoroars" from Fisher Price. Those are the toys on which she learned to suck and swallow and touch. They were her first toys that she played with. They were also her soothers. During the later part of her life she had two security items: her bunny which was named "Bud," and a dog named "Husky." She was about seven when she was given Bud by a friend at her birthday party. Kayda quickly developed an attachment to it and wouldn't sleep without him. When she was nearly ten I gave her "Husky." He got added to the necessary family members. They each had to be placed in the appropriate arm; Husky had to be in her right arm and Bud in the left. At first she was content to just have Husky and Bud while sleeping, but during the last few months of her life she needed them with her constantly. She showed this by fussing until they were where they belonged.

Kayda indicated choice by playing with what she wanted and pushing away things she didn't want. As she got older she changed what she was willing to play with. She indicated "yes" by smiling and "no" by not responding to an offered toy, video or story. This started at about the age of 9.

She clearly indicated that she anticipated TV shows. She knew what time Star Trek Deep Space 9 was on. Often I'd hear her fuss and then would look at my watch and realized that it was time for her show. She'd stop fussing the minute I turned it on and would listen carefully. She eventually seemed to know what time all Star Trek shows were on and made sure she got to watch them. There was no clock in the room that she would know about. She just seemed to know instinctively. I didn't watch the shows regularly prior to Kayda watching them. Deep Space 9 was on at the time I fed her dinner and that's the first one she started to "demand."

Kayda was able to use switches purposefully and did so without prompting. She mostly used a "Big Mack" switch which had voice messages on it. She usually had two on her tray. At one point she had a message in English on one switch and a message in French on the other. She would take the hand nearest the "French" message and reach across the tray to the "English" message, bypassing the French one. I don't remember if she showed a hand preference as in being "right or left handed". She would also reach for items on her tray and use her hands to find something that might be on her tray. Her hands were always up and reaching to touch something.

She wasn't ticklish at all (q26a). She never said any words, but did appear to understand a lot of what was said to her. Again this was pretty nebulous, it was more a sense that she understood. She clearly knew her name and would try to make sounds like I did. She never participated in play activities with other children or adults that I saw.

Kayda was skilled at calling attention to herself. She would whimper and fuss very loudly if she wasn't getting what she wanted or wasn't being listened to. Sometimes when another child was in the house visiting she'd start to whimper and when I went over to her, she gave me a big smile and would settle down right away. She appeared to be expressing jealousy and wanted to make sure she wasn't forgotten.

Kayda indicated she was in pain by crying or whimpering. She also flinched. When her hip first dislocated she kept moving that leg and flinching every time. It was like she couldn't stop trying to see if it still hurt behaving like someone pushing at a loose or sore tooth. When she was in a lot of pain she'd just lie still not moving her hands at all. Kayda smiled a lot, basically whenever she was happy. She did this when I first met her. I don't know how old she was when she started to do this.

Yes, Kayda expressed love. She started giving me "kisses" when she was just past the age of 5. I remember commenting to a nurse who had known Kayda since birth that she was doing this. The nurse looked at me and said "well Barb, you know Kayda is really low functioning and it just seems like she's doing this..." A few months later the same nurse was over and I was holding Kayda and she was "kissing" me and the nurse got all excited and said "hey she's really kissing you." This nurse started to use Kayda as a teaching case for new nurses as to why you never give up on a child or put limits on what they can do just due to a diagnosis.

Some of the milestones I would point to in her life include:

4 years 6 months: came to live with me and became an only child (previously she'd been one of many)

4 years 8 months: discovery that she breathed better lying down. Her health improved dramatically

5 years: started moving her arms slightly so that animals that previously had rested under her arms fell down

5 years 1 month: she discovered her first toy and started to chew and suck on him and anything else that was near her.

5 years 6 months: was playing with many different toys she could touch or move on her own.

5 years 7 months: traveled with us to Ontario and to Niagara Falls

6 years: started to eat small amounts of food by mouth.

6 years 5 months: her first hip dislocated and caused a great amount of pain

6 years 9 months: she had her first hip surgery and discovered her love of listening to taped stories

7 years 5 months: had her same hip dislocate again and had her second hip surgery.

8 years 2 months: her third hip surgery, this time on the other hip. She nearly died from complications after surgery.

9 years, 6 months: Make a Wish trip to Disneyland which she loved

10 years 3 months: had a serious bout of pneumonia and nearly died. That started her downhill process

11 years 1 month: became clear that she was no longer well enough to attend school or go out much

11 years 5 months: stopped tolerating food and entered palliative care

11 years 6 months: entered hospice. Died 8 days later at the age of 11 ½.

Summary of answers to the 2006 survey related to Cognitive and Awareness

81% of the children are aware of their surroundings. This is shown by facial expression (26%), vocal expression (21%), visual (20%), and body movement (19%). Others also show their awareness by auditory, hand use and other methods. 62% of the children showed this awareness during their first year. In that it appears that a child with Hydranencephaly who is less than a year is as aware as a typical child but has fewer ways of expressing this. 42% of the children are said to have changed in how aware they are of their surroundings with 97% becoming more aware and only 3% becoming less aware. This is what would be expected as a child grows and changes.

67% of the children are aware of objects. This is shown most by visual responses (30%), and hand use (23%). Other ways of showing this awareness were vocal such as laughing, cooing and fussing (11%), and smiling (4%). In this case 59% showed this awareness in their first year. In 48% of the children they showed a change in their awareness of objects with 94% of those changes being positive and only 6% became less aware.

24% are aware of their bodies. This is shown by hand use (35%), vocally (23%) and body movement (18%). Other ways of showing it are: visual (looking towards), and facial expressions such as grinning and flinching. 29% of the children showed this awareness during the first year, 24% at the age of 1 and 29% didn't specify when this started. In 47% of the children this awareness changed with all becoming more aware rather than less. Some of the specific ways children's awareness of their bodies changed are: starting to laugh when tickled, laughs have gotten louder, using hands almost compulsively at times and reactions became clearer.

91% of the children know parents and immediate family members. . This is shown in 38% of the children as facial expressions such as smiling. 23% of the children show this vocally in ways such as laughing, cooing and crying. In 16% of the children it is shown by body use in ways such as calming, turning towards and moving body. 54% of the children showed this awareness in the first year of life with 28% showing it at birth. 24% showed it over the age of 1 and in 16% of the children no age was specified. 38% increased in their awareness with all of them becoming more aware. This awareness is very similar to what is expected of a typical child. Not as many children are said to be more aware over the years but they do maintain their awareness rather than losing it as they get older.

83% of the children know the difference between a family and a stranger. 25% of the children showed this in facial expressions such as smiling or frowning. 26% showed this vocally in ways such as laughing, cooing and crying. 18% showed this visually by looking. 44% of the children showed this awareness in the first year with 15% of those showing it at birth. 24% showed it over the age of 1. The age was not specified in 32%. This awareness changed over the years in 22% of the children with all of them becoming more, not less aware.

85% of the children are able to indicate if they recognize someone or something. In 40% of the children this is shown by facial expressions such as smiling. In 24% they show this awareness vocally in ways

such as laughing or cooing. Other ways of showing this were reaching, calming, movement of eyes and reaching toward someone. In 37% of the children, this awareness started in the first year with only 15% showing this at birth. I believe this is what would be expected of any child. 25% showed this awareness at 1 and over. In 39% the age was not specified. 35% of the children changed their awareness over the years with becoming more aware and only 6% showed less awareness.

88% of the children show that they recognize familiar voices. This is done by facial expressions such as smiling in 29% of the children. 15% of the children showed this vocally by laughing, and cooing. 11% indicated this by looking at or towards people. 27% of the children showed this by turning their heads, change in eye movement and relaxing. 41% of the children showed this awareness in the first year, with 16% showing it at birth. Again, this is fairly close to what is expected in a typical child as well. This changed in 31% of the children with 93% becoming more aware and only 7% becoming less aware.

89% of the children are able to be comforted more by one person than another. In 40% of the children this was shown by calming with one person. 29% showed it vocally by crying. 24% showed it by cuddling, snuggling, relaxing and being comfortable with one person. 59% of the children showed this preference in their first year, with 42% of those showing this awareness at birth. 12% showed it over the age of 1 and in 28% no age was given. Only 19% of the children had a change in this awareness over time. Of the 9 children specified in the survey 6 (66%) became more aware and 3 (33%) became less aware.

67% of the children show fear or dislike of something or someone. 52% of the children show this primarily by vocal expressions such as crying or fussing. 20% of the children show this by body use such as startling. In one incidence a child has reacted to fear or dislike by having a seizure. 38% of the children showed this in the first year with 16% of those showing it at birth. In 19% of the children this has changed over time. In 87.5% of the children this was a positive change. in 12.5 % (1 child) this behaviour happened less.

45% of the children show a strong aversion to someone or some thing. The responses to how this is shown were too varied to fit into a table. Some of the things children had a strong reaction to are nebulizer masks, certain people and smells, cold water, loud noises, parts of videos, and many more. One child was reported to avoid lying on a wet sheet or pillow by turning himself around so his face isn't on the wet spot. Many of these reactions are those we'd find in a typical child of the same age. 41% of the children showed these reactions in the first year of life. 39% over the age of 1 and in 20% no age was given. 24% of the children showed a change in what they disliked or in how it was shown. 83% of the children are reported to react more or change what they dislike while 17% (1 child) reacted less over time.

41% of the children are said to get mad or have temper tantrums. The responses were too varied to create a table of common words. Some of the responses include: "when he's tired or has had enough", "when you change her", "if she wants to be held and you can't pick her up right away." These are all situations in what any child may respond. In 33% of the children this has changed with 70% of the children either getting upset more or increasing the number of things they react to in this way. In 30% of the children they react less.

43% of the children have a favorite toy. This is shown by facial expression such as smiling or grinning in 30% of the children. It was shown vocally by laughing, crying and making sounds in 27%. 20% of the children showed this with hand use such as holding and pushing of the toy. This changed over time in 33% of the children with 73% having positive changes and 37% had a decrease in this behaviour.

62% of the children have a favorite activity. These include: watching videos and listening to stories, eating, playing with toys, bath time and many more. This has changed in only 14% in a variety of ways such as "likes physio", and "awareness of them has increased" In 2 of the children their like of a particular activity decreased.

51% of the children have a favorite position or piece of equipment. There is a wide range of their favorites. These include: standers (a contraption that allows a child to stand), lying down, being on mom's lap, lying on her side and being in a recliner. A lot of the children's favorite position is being held by mom or dad. Very much like any child. 29% of the children show this with facial expressions as smiling. 24% of them show this vocally by laughing, crying, and cooing. 29% include reactions such as the child being happy or happier, relaxing, and being comfortable. This has changed in 22% of the children. 50% of the children are reported to increase what they like and 50% changed by liking fewer things.

40% of the children have a security item. These range from a stuffed toy, favorite blanket, a favorite position on mom, mom and many more. The children all show this attachment in different ways. In 11% of the children it's shown in facial expression, vocally, visually and in body use. Some of the ways it's shown are smiles, grinning, laughing, crying, holding, and calming. In 53% of the children other words used to express the attachment are: relax, comfortable, rest, hug, content and quiet. These responses are those we'd see in any child. In 40% of the children this attachment has changed with increasing their attachment and in 60% the attachment has become less.

27% of the children can make choices. The responses to the question as to how it is shown vary widely. They include with switches, eye gaze, closing lips if he doesn't want something, blinks for yes, stares for no, rolling eyes for another choice and many more. 61% have changed in how they make a choice. In 93% of the children there was an increase or change in how they indicated a choice or an increase in ability to make a choice. In 7% of the children they "used to" play with a particular toy. A few (not included in the total) are said to make fewer choices as they've grown older.

15% of the children have a "yes" "no" response. This is shown in a wide variety of ways. Some of them are: smiling for "yes" and no response for "no", smiles and blinks, ignores you if it's no and "uh huh" or "no". In 32% of the children this response has changed. There were only 2 common words used, in one case the child's choices improved and in the other it became less.

37% of the children anticipate or expect events such as tv shows, going to familiar places or other daily events. The events the children anticipate varies widely. They include anticipating meeting familiar people, getting tickled, expecting a particular tv show, going to daycare, expecting the dog to greet him in the morning and many more. The children show this in a wide variety of ways. 14% show this by facial expressions such as smiling or grinning. 36% of the children show this vocally in ways such as laughing, crying or fussing. 7% of the children show this by kicking. In 32% other words were used such as happy, tense, excited and eyes. 26% of the children showed this during their first year and 58% showed it at 1 or over. This would be expected in a typical child as well. In 14% of the children this anticipation was changed as they got older. In 80% of the children this anticipation increased and in 20% the anticipation decreased.

38% of the children are said to understand what is said to him/her. In 50% of the children this was shown by smiling. Other ways it's shown are cooing (10%), looking (10%), holding feet still (10%), eyes moving (10%) and listening (10%). The ways this is shown vary widely. They range from "opens mouth for toothbrush", "lifts foot for sock", "smiles in response", "stop an activity when asked to" and many more.

67% of the children can call attention to themselves. In 98% they do this vocally by crying, yelling, laughing, cooing, hollering, making noises and shouting. The children would do this in a wide variety of situations. Just as is done in a typical child.

96% of the children can feel pain. This is shown by facial expressions such as wincing, grimacing and flinching in 14%, vocally in ways such as crying, screaming and yelling in 78%, and body use such as wriggling, pulling away and startling in 4% of the children. 4% of the children are said to have seizures in response to pain. The majority of the families are told that there is no way their child can feel pain. The responses to this question show that this is definitely not the case and those children with Hydranencephaly show pain in the same ways any child does.

89% of the children smile. It happens at any time a child is happy such as on hearing the voices of family members, when told what is going to happen, the sounds of toys and many more. In 34% of the children their smiling has changed over the years. 86% of the children whose smile has changed report that their child is smiling more or it has improved. In only 14% the children are said to smile less. 65% of the children started to smile before the age of 1 with 9% reported to have been smiling at birth. 18% started at the age of 1 or older and in 16% no age is given.

The following questions are out of sequence on the survey. They also are more dependent on hand use and the ability to control parts of the body. But they also show to a certain extent evidence that the child is aware and interacting with their environment.

31% of the children can activate switches in a purposeful way. This is 27 out of 88 people who responded. 38% (16 out of 42 participants) answered yes to the question "If yes, does your child do so without prompting. The numbers of the 2nd part of the question really don't match as only 27 of the children are said to activate switches.

26% (23 out of 90 participants) of the children will reach for objects. 22% (15 out of 67) will do so spontaneously without prompting from others. Again, the numbers really don't match.

9% (8 out of 90) of the children have been seen to turn towards or rub an area of the body that hurts.

56% of the children are said to be ticklish. It is shown by facial expressions such as smiling in 19% of the children. 57% of the children show it vocally such as laughing, crying, cooing and giggling. 24% show it by body movements such as kicking, wiggling, moving, squirming, and pulling away. 38% of the children will react if you take the child's hand and move it against the child's body in the same way you do when you do the tickling. In 22% of the children the reaction is the same as when the adult does the tickling. This is shown by squirming, twitches and cries in the same way, getting very giggly and laughing.

14% say and use appropriately certain words. The words are quite varied. Most of the children seem to use a few common words such as mom, no. A few of the children use a large number of words. 34% of the children understand specific words. 21% understand mom, mama, etc., dad, papa in 3%, their name in 28%, no in 28%, yes in 10%, and bottle in 10%.

16% of the children have a communication device. These include a Big Mack for 60%, Eye gaze in 7%, computer system in 7%, and an unspecified device in 27%.

66% of the children respond selectively to their names. 29% of the children can echo or imitate when they're spoken to.

16% of the children will take turns in play activities. 31% will build up play sequences by getting more excited as the game progresses. 34% will show signs of "wanting more" during play or joint activities.

Summary

The purpose of this study was to get a clearer picture of whether or not a child with Hydranencephaly was truly aware of their surroundings and to find out if they could learn. This was accomplished by a survey asking questions about all aspects of having a child with Hydranencephaly. There was a section in that large survey which asked detailed questions about awareness and responses. Each set of questions included a yes no question on each aspect, open ended questions giving a description of how the child showed this and describing change, a question on the age a child was when they started to show a particular reaction and a yes no question about if the child's awareness had changed. 106 families completed this survey although not all participants answered all questions.

Is a child with Hydranencephaly aware of his/her surroundings?

According to the survey results, we know that a child with Hydranencephaly can feel pain, know their parents and immediate family, express themselves by smiling, are comforted by one person over another and recognize familiar voices. Some other areas that are represented include: have a favourite toy, make choices, and reach for objects.

According to the percentages shown in the previous section parents report that the overwhelming majority of the children feel pain, know parents and immediate family, smile, are comforted more by one person over another and recognize familiar voices.

Can a child with Hydranencephaly learn?

Yes. In a majority of the areas of awareness a significant number of children changed in their reactions and awareness over the years with the vast majority increasing in this awareness or interaction. By changing in this way the child shows that there is a progression in what they know or in how they react in a particular situation.

How would you know if a child with Hydranencephaly is learning?

By an increase in the particular activity or awareness. They mostly show awareness with facial expressions or vocally. Other ways we can tell if a child with Hydranencephaly is aware is by body movement, using their hands, and calming. Their clues are much more subtle than in a typical child but can be read by those who are familiar with them.

From what has been seen in this survey, having someone who knows a child well is vital in knowing if the child is aware. The children each show their awareness in a unique way so part of the communication is having a person to interpret it correctly. Another issue is that the children respond to many things in a similar way such as vocally or with facial expressions. This means that when a child smiles it could be because he likes something, recognizes something or someone or many other possible reasons. Again, having someone who knows the child well is essential in them sharing their awareness. This is likely why doctors or other professionals who see a child for short periods of time think that they have no awareness or ability to learn. It takes time for someone to learn to read each child's cues.

I don't believe we can develop an "ages and stages" cognitive assessment for a child with Hydranencephaly but we can certainly say to a new parent that their child will be aware and learn to interact with his/her surroundings. And, we can say without doubt that our children feel pain.

Appendix 1

Summary of cognitive development and awareness

Skill/reaction	percent	Shown most by	shown in first year	Change	Positive change
Feel pain	96%	Vocal (78%)			
Know parents and immediate family members	91%	Facial expression (38%)	54%	38%	100%
Smile	89%		65%	34%	86%
Comforted more by one person over others	89%	Body use (calming) (40%)	59%	19%	66%
Recognize familiar voices	88%	Facial expression (29%)	41%	31%	93%
Recognize someone or something	85%	Facial expression (40%)	37%	35%	96%
Know difference between family members and strangers	83%	Vocal (26%)	44%	22%	100%
Aware of surroundings	81%	Facial expression (26%)	62%	42%	97%
Aware of objects	67%	Visual (30%)	59%	48%	94%
Show fear or dislike	67%	Vocal (52%)	38%	19%	87.5%
Call attention to themselves	67%	Vocal (98%)			
Respond to their names	66%				
favorite activity	62%			14%	
Ticklish	56%	Vocal (57%)			
Favorite position or piece of equipment	51%	Facial expression (29%)		22%	50%
Show a strong aversion	45%		41%	24%	83%
Have a favorite toy	43%	Facial expression (30%)		33%	73%
Get mad or have a temper tantrum	41%			33%	70%
Have a security item	40%	Mixed (57%)		22%	50%
Understand what is said	38%	Facial expression (50%)			
Anticipate or expect events	37%	Vocal (36%)	26%	14%	80%
Activate switches	31%				
Echo or imitate what is said	29%				
Can make choices	27%			61%	93%
Reach for objects	26%				
Aware of their bodies	24%	Hand use (35%)	29%	47%	100%
Have a communication device	16%				
Take part in play activities	16%				
"yes" "no response	15%			32%	50%
Say & use words	14%				
Turn to or rub a body part that hurts	9%				

Appendix 2

Questions as asked:

1. Is the child aware of his/her surroundings. How is this shown? At what age did this awareness start? Has this changed over the years? If yes, how?
2. Does your child show an awareness of objects? How is this awareness shown? Has this changed? How was this shown?
3. Does your child show an awareness of his/her own body? How is this expressed? At what age did this awareness start Has this changed over time? How has this been shown?
4. Does your child know you and your immediate family members? How is this shown? At what age did this awareness start? Has this changed over time? How has this awareness changed?
5. Does your child know the difference between a family member and a stranger? How is this awareness shown? At what age did this awareness start? Has this changed over the years? How has this changed?
6. Does your child indicate if he/she recognizes something or someone? How is this awareness shown? : At what age did this awareness start? Has this changed over time? How has it changed?
7. Does your child show that he/she recognizes certain voices? How is this shown? At what age did this awareness start? : Has this changed over time? How has this changed?
8. Can your child be comforted more by one person than another? How is this shown? At what age did this start? : Did this change over time? How did it change?
9. Does your child show any indication of fear or dislike? How is this shown? At what age did this start? Has this changed over time? How has it changed?
10. Does your child show a strong aversion to someone or something (something they don't like to look at, particular activities/stories, smells, people, etc)? What? At what age did this start? Has this changed over time? How has it changed?
11. Does your child get mad or have temper tantrums? Under what circumstances? has this changed over time How has it changed?
12. Does your child have a favorite toy? What is it? How does he/she show it? Has this changed over time?
13. Does your child have a favorite activity? What is it? Has this changed over time? How has it changed?
14. Does your child have a favorite position or piece of equipment? What is it? How does he/she show it? Has this changed over time? How has it changed?
15. Does your child have a security item? What is it? How is it shown? Has this changed over time? How?

16. Can your child make choices? How does he/she indicate a choice? Has this changed over time? How has it changed?
17. Does your child have a yes/no response? What is it? Has it changed over time? How has it changed?
18. Does your child anticipate or expect events (ie; a particular television program, going to visit someone familiar, other events of daily routine, etc)? To what? How is this shown? At what age did this start? Has this changed over time? How has it changed?
19. Does your child understand what is said to him/her? How is this shown?
20. Can your child call attention to his/herself? How? what situations would he/she be most likely to do this?
21. Does your child feel pain? How is this shown?
22. Does your child smile? In what situations? Has this changed over time? How has it changed? At what age did your child start to smile?
23. Can your child activate switches purposefully? If yes, does he/she do this without prompting?
24. Does your child reach for objects? : Does your child do so spontaneously, when the object was not presented by others?
25. Have you ever seen your child turn to or touch or rub a part of his or her body that hurt?
26. Is your child ticklish? If yes, how does he/she show this? If your child is ticklish, and you take the child's hand and move it against the child's body in the same way you do when you do the tickling, will he/she react? Is the reaction the same as when you yourself do the tickling directly? If yes, how
27. Does your child say and use meaningfully any words? What words
28. Does your child understand any specific words? What are they?
29. Does your child have a communication device? If yes, what kind?
30. Does your child respond selectively to his/her own name?
31. Will your child echo or imitate you?
32. Will your child take turns with you in play activities?
33. Are you able to build up play sequences in which your child gets more and more excited as you keep at it?

34. Does your child show signs of "wanting more" during play or other joint activities? How is this expressed?

References:

2006 Hydranencephaly Survey