General Overview of Children with Hydranencephaly

According to a parental survey conducted from 2005-2007

Hydranencephaly Information Network

2006 survey summary

Introduction

As the years have gone by, the families of children with Hydranencephaly have wanted to know more about this condition and what is and isn't common to our children. Since 2001, 5 informal surveys have been conducted. Each has had a variety of questions and numbers of people who participated. In 2006, a much larger/longer survey was created. Because of the volume of questions asked and answered it has taken a long time to compile the results of this survey. This document will give a summary of what has been learned from our children based on this survey. I am a parent, not a medical professional so this will be presented informally and is intended to mostly be read by parents and lay people who may have an interest in Hydranencephaly.

Summary of the responses for Kayda

Kayda was born in Vancouver BC (Canada). Her mother was Caucasian, father unknown. She was missing all of her cerebral hemispheres except for a small amount of occipital lobe and basal ganglia. Her skull was full of fluid. At the time this survey was filled out, she was deceased. She was born on December 2, 1988 and died on June 23, 2000. She was 11 ½ years old when she died.

The primary cause of her death was a gradual deterioration. She started to have trouble digesting her food and became generally uncomfortable just over a year before she died. A lot of the problems seemed to have started after a very bad pneumonia in March 1998. Gradually she was able to do less and less. She started having trouble swallowing saliva and needed suctioning. And, she started to sleep most of the time. Most of her last 3 months were spent sleeping in her new "comfy" chair (a custom made chair).

Kayda was first born. There was little knowledge of her biological family so it's not known if there were any family members who had other neurological disorders. Kayda was put up for adoption at birth and had a variety of homes in her first few years including a fair amount of time in a long term care facility in their Neonatal Abstinence Syndrome program. She was placed in my home (similar to a foster home) at the age of 4 ½. She remained with me until her death.

Her Hydranencephaly was diagnosed during her first day of life via a CT scan. I was told that the CT scan was done as her "head looked odd". Her birth mom was 16 years old and had no prenatal care. Suspected cause of her condition was prenatal drug exposure. She had no additional health problems at the time of her birth.

Health Issues

Kayda had allergies. She was allergic to Latex and Medications. Her latex allergy was acquired throughout her life due to multiple surgeries and a latex catheter left in for several weeks. Once it was diagnosed it was a severe life threatening allergy. Although she was thought to be allergic to a particular class of antibiotics, her main difficulty was with seizure medications. There were very few of the many that were tried that she could tolerate.

Kayda had hydrocephalus which was apparent at 2-4 weeks of age. She received a Ventricular Peritoneal (VP) shunt. She needed 1 revision to it about a month after the first was placed. She had no further problems with her shunt and had it the rest of her life. We actually forgot she even had a shunt it worked so well. She also had microcephaly which is a smaller than normal head. That was due to the fact that she had little brain tissue other than at the back. The top of her head was somewhat pointed which also was due to the lack of brain tissue. Her hair covered that up.

Her most common health problems were Neurological, Feeding and respiratory issues. She didn't have much trouble with spasticity. She had Asthma/Reactive Airways disease. It was usually most prevalent with a cold virus. She did take a preventative medication (Pulmicort) via a nebulizer twice every day. When ill she'd receive Ventolin as well. When ill she'd get manual chest physio and postural drainage 4 times a day. As she got older she didn't tolerate the chest physio well so we moved to a vibrator. Eventually she stopped tolerating that so I didn't do chest physio on her. I mainly just changed her position frequently. We did what I called "mommy chest physio" I sat her on my lap with her head on my shoulder and her arms around my neck. That opened her chest and got her in a good position for breathing and coughing.

She never had a tracheostomy nor did she ever use a ventilator or Bipap machine. The Bipap wasn't widely used during her life and no one would consider putting her on a ventilator for an illness. She did, however use a ventilator during surgeries. She always came off of it fairly quickly. She didn't have sleep apnea. Kayda was on oxygen at home from about the age of 9. During her last year she was on it all the time. Up until a few months before she died she was only suctioned when ill. The last few months she needed help with clearing saliva as she could no longer swallow well. She had pneumonia numerous times. No one was ever certain whether it was aspiration or bacterial or viral. Often when she was sick it was assumed that it was aspiration pneumonia but often just turned out to be a bad cold. She had numerous bouts of tonsillitis and ear infections.

Although it was suspected through much of her life, Kayda was only diagnosed with Diabetes Insipidus after her 3rd hip surgery at the age of 8. At first she needed the DDAVP but was very sensitive to that so we just controlled her sodium level with lots of fluids (3-4 litres a day). She had no further endocrine problems.

She did have premature puberty. It started at about the age of 3. She started menstruating at the age of 10 years 3 months (right at the same time as she had a bad pneumonia and started being very uncomfortable). She didn't bleed for more than a few months but I could always tell when it was that time of the month as she'd have a 16 hour non convulsive seizure. It would start early in the morning with her being extra sleepy and very cold (lower than 35 Celsius). It would end around 3 am and she couldn't understand why I wasn't interested in partying with her once she woke up. I always stayed with her during these episodes just in case it developed into something else. In addition to the above seizures her over all seizures increased drastically each month. She went from having 2 or 3 small focal seizures a day to hundreds every day.

Kayda had no history of urinary tract infections or any other problems of that type. She definitely had trouble controlling her temperature. It was most often very cold (34c or lower). When it was super hot out it was difficult to keep it down though. She also had trouble sleeping. Yikes, it was bad. We tried just about everything to help her to sleep. Choral hydrate was the first med we tried and it sedated her so much I couldn't tell if she was still breathing. She'd also holler most of the following day. Then we tried Melatonin but it didn't work either. The dr said that as melatonin is processed in the hypo thalamus it was likely she was missing all or part of hers. I also tried Calmes Forte and herbal tea. Neither worked. I remember once when Kayda had been in the hospital for awhile. I saw her paediatrician downstairs and told him I wanted to talk to him. I can still remember him backing away from me saying he didn't know what to do to get Kayda to sleep. She would often sleep only every 2nd or 3rd night and would shriek loudly day and night in between. Finally, after a suggestion on an email group it was suggested to try Trazodone. The dr agreed and it was wonderful from day 1. It's an antidepressant with a sedative effect. She didn't have one sleepless night for the rest of her life.

Kayda had trouble with excess saliva. It got a lot worse after starting the seizure med Vigabatrin. Again, after a suggestion from another family I asked the dr if she could try Robinul. It worked well for her. The excess saliva continued after stopping the medication so she stayed on Robinul for the rest of her life. It had to be increased a few times. We also tried the patch but it didn't work at all.

Constipation was also a problem. We tried multiple meds as well as natural remedies such as "poop goop" (a mixture of a variety of fruit mixed with senna powder). None worked very well. Although I'd been determined to make sure she didn't become dependent on suppositories or laxatives she did. A dulcolax suppository and Magnolax worked best for her. Agarol wasn't bad either. She had no med port or venous access device. It would have been nice as it was so hard to get an iv in or do blood work but it was never suggested for her.

Seizures

Yes, she did drift into absences. These were her primary type of seizure. They happened numerous times each day especially after she started her period. They'd last 5-20 seconds. We always knew when she was back as she'd start to look around and interact again. Kayda definitely had seizures although none were ever picked up on an eeg. It wasn't until near the end of her life that it was ever suggested that she was having brain stem seizures which aren't usually picked up by a eeg. Sometimes she could be brought out of the absence style seizures by running your hand over her eyes. It was touching to watch the children at school just automatically do that for Kayda. Although I answered yes to the question as to if something could bring on a seizure I'm not sure about that.

We mostly tried conventional medications for her. Very few either worked or were tolerated by her. She reacted badly to most. Nitrazapam, Vigabatrin, and Lamontrigine were the only medications she could take without major side effects. None of the other treatments (Ketogenic died, Vagal nerve stimulator) were offered for her. During the last few months of her life her seizures increased drastically to where it was thought she was always seizuring by medical people who saw her. We were going to try the

Ketogenic diet and she started the testing and such that would be needed before she could try it but it soon became clear that she was dying so we stopped.

She did startle easily. She went through a stage when she was about 9 years old where the tiniest noise would make her jump and then she'd have this long bout of difficulty breathing and general upset. This would go on until I'd picked her up. Her school people thought they were seizures but the doctors had seen her and said no they weren't. They stopped right around when we were trying Tegretol I think. The side effects from that overly sedated her and once she was off of it there were no more extreme startling episodes. We never knew why she had them and then stopped. I wondered if it was because she was getting more alert and couldn't handle all the input her system was getting.

Kayda had 5 different types of seizures or behaviour that was thought to be a seizure.

1. Shiver type seizures, momentary, started at about age 8, controlled by Vigabatrin
2. Staring spell or fixed focal seizures; lasted 5-20 seconds, started at around age 9, at first she only had a few a week, by the time her period started at age 10 she was having hundreds a day, last year of her life she was having them almost constantly
3. Full body tremors: started at age 11 shortly after starting to take Topomax, only happened while deeply asleep, would last until she woke up-several hours. They stopped happening within a couple of days of stopping the Topomax.
4. Spasm type seizures, these were never confirmed as seizures but could have been, they were position dependent and involved grimacing and tightening one side of her body
5. Coma type, non convulsive seizures, had a number when young and then started to have them monthly with her period, they would last exactly 16 hrs

Feeding

Kayda received a feeding tube at about 18 months of age. For several years while living with me she was able to eat both by tube and by mouth. By chewing and sucking on her toys she taught herself to suck, chew, swallow and cough. Medical and other professionals were very nervous about her eating orally as they just assumed she would aspirate as earlier in her life she had aspirated a lot. During those years of eating orally she never once aspirated. But, as time went on she wasn't enjoying eating enough for it to be worth the risk. When eating she only ate smooth purees. Her favorite food was pea soup. We think she had mild reflux through her years but it didn't become an issue until she was 10 or older. Reflux and poor motility (how fast the food goes through the system) became a major problem during the last year of her life. She tolerated less and less food at lower and lower rates during that time. She never had the Nissen Fundoplication (where the top of the stomach is tightened to prevent food from coming back up). She wasn't healthy enough for it by the time reflux became an issue. We also considered going to a jejunostomy tube which helps with reflux, but again, by that time it was clear she was dying and it was decided not to do any more procedures that required an anaesthetic or sedation.

Physical

Kayda had mixed tone; both floppy and tight. She did have clonus which is a tremor of the leg not related to a seizure. Both her hips became subluxed (partly out of joint) and eventually both dislocated. She had 3 hip surgeries. Two were the full reconstruction of the hip joint and the other was called an open reduction where she was opened up and the hip was put back into place). She wore an abduction cast called a Petrie cast which were just on her legs with a bar in-between. Once the casts came off she wore a Scottish rite abduction brace 24 hours a day at first and then just at night. She was in casts or a brace for over 2 years. But the relief from the pain of the dislocated hips was well worth the discomfort and difficulties of the surgeries.

She was diagnosed with a mild scoliosis at the age of 5. She started wearing a TLSO (back brace) and there was no change to her scoliosis throughout the rest of her life. It was hard for the Orthotist to find a brace that fit and was comfortable for Kayda as she grew. She also had a Kyphosis (forward hunch of her back). The brace helped that not to progress as well. She developed contractions (a tightness that can't be stretched out) in her hamstrings (tendons at back of legs). They'd hope do to a release of her tendons during her last hip surgery but felt she was able to tolerate extra time anesthetised. Under advice from several Physio Therapists I started putting her in her stander daily and used long leg splints at night. With this combination the tightness in her left leg went from a -40 degrees to a full extension in only a few months. In addition to the back brace and abduction brace she also wore AFOs (ankle foot orthotics) during the day and hand splints at night. These prevented further deformities. She used a standard wheelchair. We had a ceiling track lift system for her which was wonderful. It made caring for her so much easier. The best thing was that it went from over her bed, through the door frame and into the bathroom ending over the bath tub. It made bathing her easy. She got a custom lounge type chair (called a Comfy Chair) just a few months before she died. It made those last months more comfortable for her. In earlier years we used both a Feeder seat and a bath seat as alternate positioning devices. She slept in a manual hospital bed. We transported her in her wheelchair in a van with tie downs.

She started rolling at about 5 years of age. She mostly rolled from her back to either side. I don't remember if she ever got all the way over. If she did, it didn't happen often. She stopped rolling shortly after her first hip surgery. The surgery realigned her hips so that her legs were further apart so rolling was no longer possible. Kayda moved her hands, arms and fingers. Her hands were always up looking for things to explore and pick up. She didn't often pick up items though. She mostly just touched them. She could bring her hands to midline with no problem. She started this at about 5 ½ years of age. She also moved her legs. She didn't kick or bicycle with them though. She never walked or sat up either unassisted or when supported by her legs only. She never crawled. She did move on the floor by kicking her legs when she was younger. This stopped after her hip surgeries.

Social and Emotional

Kayda did appear to give hugs and kisses. She could definitely cry. She was very fussy as a baby but rarely fussed when she was older. She was happy most of the time.

Vision/Hearing

She could see some of the time. Testing at the age of about 7 showed that she could see up to 5 feet away. She saw light objects in a dark room the best. Her vision wasn't very consistent. I did see her peer around me once when the weather man was on tv. She definitely responded to lights being turned off and on. She showed this by her eyes stopping flickering. She'd turn both her head and eyes towards bright objects. One day she very clearly noticed Christmas lights on the tree that had been put up during the time she was out of the room. She had nystagmus (repetitive jerking movements). It would stop when she was listening to something or recognized something. She was diagnosed as having cortical visual impairment. She never was diagnosed with optic nerve hypoplasia.

She heard very well. She would turn both her head and eyes towards the sound of my voice. She would listen closely to things she wanted to hear (certain stories) but fuss if she didn't want to hear them (the "wrong" type of story or music).

Kayda definitely made sounds. She never said any words. It appeared that she understood most of what was said to her but this was mostly a feeling. I always used the same words as part of her daily routine so I think she understood those words. She understood Mom, mist, story, would smile if I said "hocapontas" but remain stony face if I said "Pocahontas"). She clearly responded to her name. She also echoed me. She didn't engage in play activities.

Kayda went to school and was fully included with her age appropriate peers. This worked fairly well for her up until the end of grade 4. Then she got more restless and noisy and it just wasn't working. So, in September for grade 5, although she was registered in grade 5 she was in a grade 1-2 split class. This worked very well for her. We did have to get her to listen to younger stories than what she preferred at home. Her principal was brilliant at making sure Kayda was part of the school. She stopped going to school part way through grade 5 as she started getting sick every time she was taken out.

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.

General Summary of the results of the 2006 survey

Demographic information:

Gender of children was 57% males and 43% females. 79% of the families participating in this survey were from the US. There were 7% from both Canada and the UK. There were also families from Germany, Australia, New Zealand, Thailand and Puerto Rico.

A question was asked about ethnicity of parents but these numbers weren't significant so are not included in this summary.

Birth order of child born with Hydranencephaly didn't seem to be specific as the numbers were similar for each category. See table below.

	Number	Percent
Only child	27	28%
1st born	21	21%
2nd born	23	23%
3rd or later born	27	28%
Total	98	100%

A series of questions were asked about incidence of other people in the family who had either Hydranencephaly or another serious neurological problem. One family had another family member with Hydranencephaly. A small number of families had other family members with a severe neurological condition (18%). So it would appear that there is somewhat of a higher risk of having a child with a

severe neurological condition in families of children with Hydranencephaly. But, in our support group no family has had a second child with Hydranencephaly.

Age of mother at time of birth.
In this survey 38% of the mothers were under the age of 20, 35% between 20 & 25, 17% from 26-30 year olds and 11% were born to mothers over the age of 30.

As included in the 5 survey comparison that was created in 2011 by Barb Aleman and is currently online at www.hydranencephaly.com/researchresults.htm:

"Our numbers on mother's age have gotten the attention of Marc Lubinsky as although the percentage of mothers who were over the age of 20 is greater, the higher numbers of mothers under the age of 20 is higher than what would be expected statistically. He is the author of 2 articles online which can be read at: http://www.hydranencephaly.com/causeofhydran.htm. They have been on both our website and in the books. I had discounted his results due to the fact that there were a higher percentage of mothers over 20. But as he pointed out: "*there is no question that there is an increased rate in younger mothers. Mothers under 20 make up about 10% of all births, so they would be expected to have roughly the same % of children with Hydranencephaly- instead, the sample from your survey had 41%, clearly an increase! Also, of all teenage births, about 1/3 are to women under 18 years old, but those women had 73% of all children with Hydranencephaly born to teenagers! In other words, the effect is greater as mothers get younger*." "

There also doesn't seem to be a correlation between time when mothers started receiving prenatal care and those that didn't. 32% had no prenatal care, and 34% had it during their first trimester.

Cause of Hydranencephaly
The 2 most common causes of Hydranencephaly are prenatal stroke (47%) and Unknown (37%). Other causes include: prenatal drug exposure (6%), prenatal infection (5%), death of twin in utero (4%), and 2% were caused by an illness after birth. According to the definition of Hydranencephaly which appeas on the home page of the most complete resource on Hydranencephaly (www.hydranencephaly.com) : The damage to the brain usually occurs in the 2nd or 3rd trimester of pregnancy and can occur up to a year after birth as well. This definition was partly written by Dr Bjorn Merker who has written in several journals about children with Hydranencephaly.

14% were said to have other severe health issues at birth including Spina bifida, kidney reflux, chiari malformation and a few others.

Health issues in children with Hydranencephaly
Allergies
64% of the children were said to have allergies. These included latex, environmental, medications and food. About the same number had environmental, medications and food allergies.

Hydrocephalus
78% of the children whose parents participated in this survey had hydrocephalus. 27% of the children had their hydrocephalus diagnosed at birth. 22% before birth, first month 20%, 17% at 1-3 months and 6% after the age of 6 months.

94% had surgery for their Hydrocephalus. All had the Ventricular Peritoneal Shunt. 96% still had a shunt at the time the survey was filled out. 49% had their shunt placed at 1-3 months. 66% have needed to have their shunt revised or replaced with the highest percent (40%) replaced at 1-6 months after the first was placed. Some children (34% in this survey) never need a shunt revision or have 1 and never need another one. We know of one person who had the same shunt until the age of 16 and then started needing new shunts frequently. Most of the children do well long term with one shunt.

24% are said to have Microcephaly (smaller than normal size head).

Most common health concerns

	Number	percent
Respiratory	48	47%
Neurological	63	61%
Feeding	60	58%
Spasticity	51	50%
Other	12	12%

As you can see the children have a wide number of health problems with each child often having more than one type of problem.

Respiratory problems
66% of the children are said to have respiratory problems. There are a wide variety of conditions and treatments for the children.

71% have Asthma or reactive airways disease. Often the asthma is caused by something specific as aspiration, or a cold virus. There are a variety of treatments available for families to try. Most commonly used (28%) are aerosol or nebulizer medications. Another common treatment (18%) is manual chest physio. Increasing numbers of children are getting their chest physio via a “vest” which is a jacket that is worn that shakes them gently. Only 8% were using it when this survey was conducted. I’m expecting that number to be higher now.

Another respiratory treatment that is being used more commonly in children is the use of a Tracheostomy. When our group started in 1998, only 2 of the first few children we knew of had tracheostomies. And, their parents had had to fight very hard to convince doctors to do this procedure in the children. In the 2006 survey 9 (9%) of the children had a tracheostomy. In the 2010 survey 13 of the children had them. Another progression in treatment of our children is the use of a ventilator or Bipap. In 2006 18 (40% of those who answered question) children had used either a ventilator or Bipap

to aid in breathing at one time or another. 13 (31%) were still using them at the time of the questionnaire. In 2010 32 used a Bipap and 14 used a ventilator. This is a huge improvement in care for the children. In all of the children I know of who have used either their quality of life has greatly improved. It's the same in those who have tracheostomies. None of these treatments have been the horrible, invasive procedures as they were claimed to be by doctors in previous years.

Sleep apnea is now being diagnosed more frequently in children with Hydranencephaly. In 2006 28 children were diagnosed with this condition. Only 4 (13%) used a CPAP (continuous positive air pressure) which is usually the treatment for sleep apnea. These questions weren't asked in the 2010 survey.

32 (46%) use oxygen at home and 50 (88%) need suctioning. Despite the claim by doctors that all children who have Hydranencephaly will get pneumonia and die from it, only 43 (46%) have had bacterial or viral pneumonia, and 33 (35%) have had aspiration pneumonia. That's well under half of the children who have had these illnesses.

Other health issues in a child with Hydranencephaly

16 (26%) are reported to have Diabetes Insipidus which is a problem with regulating urine output and thus causing an elevated or decreased sodium level. It can be managed by medication, or fluid intake.

Hypothyroidism in 7 (13%), other endocrine problems in 5 (5%) of the children.

Approximately 81% have what is considered to be early puberty (Precocious Puberty). This is described as the onset of puberty before the age of 7-8 in girls and 9 in boys.

5 (6%) of the children are diagnosed as having a Neurogenic bladder. This is a problem in which a person lacks bladder control due to a brain or nerve condition.

Temperature control is a difficulty for the majority of the children 76 (78%) with 65% reporting a tendency to low temperature, 11% to a high temperature and 24% reported their children had both high and low temperatures.

59% were reported to have difficulty sleeping. A large variety of sleep medications were tried. Some helped some children and not others. The only real connection between what was tried and what worked was Melatonin. This was tried in 20 of the children and it worked in 2 of the children.

This finding along with some of the above categories (temperature control, and puberty) ties in with the type of damage to the brain often seen in a child with Hydranencephaly. Not all, but it seems to be connected in a lot of the children. Dr Bjorn Merker has started to draw a connection between those who had a particular artery blocked in a prenatal stroke and these difficulties the children face and which areas of the brain stem are damaged or missing.

Another common problem is excess saliva/secretions. This is a problem in 63 (80%) of the children. Of the medications tried, Robinul (glycopyrrolate) has the best results with 14 out of the 17 who tried it reporting that it worked the best.

In 2006 15 (18%) of the children had a med port or other permanent access device through which to give iv fluids and medications. As children with Hydranencephaly often have tiny veins that have been accessed many times and so it is difficult to put in an iv, these devices are very good to have.

Seizures in a child with Hydranencephaly

Many parents are told that their child does not have enough brain to have seizures. As parents observing out children, we often are convinced that he/she does have them (89%). The big problem in diagnosing seizures in our children is that diagnostic tests and treatments are designed for children with a full cortex. Therefore many of the children who parents and doctors are convinced are having seizures never have them show up on an EEG. Only 42% are picked up on the EEG which is less than half the children thought to be having seizures. The type of seizure reported by parents varies widely. The specific seizure descriptions can be obtained upon request to Barb (angelbearmom@shaw.ca) Many families describe a number of different seizures seen in their child. Some of the children (64% sometimes, 6% always) can be brought out of a seizure by an external touch or stimulus. Others are said to have seizures brought on by a external event (touch, sound, pain, stress, etc). 49% reported this for their child.

A wide variety of treatments were tried in our children. These include Conventional seizure medications/treatments, herbal or alternative medications/treatment, Ketogenic diet and the Vagal nerve stimulator. Conventional treatments worked in 77 children. The Ketogenic diet and Vagal nerve stimulator are less frequently used treatments. 3 families have tried the Ketogenic diet and it's worked for 2 of the children. Vagal nerve stimulator has been tried in 1 child and it worked well. In the years since this survey was created there are more children using these methods with good results. It will be interesting to see over a period of years how often these treatments are used and how well they work for our children. A factor in many children not responding well to medication is that the medications are designed for cortical seizures and our children's seizures mostly originate in the brainstem just like in whether seizures are seen on EEGs.

Something that may or may not actually be a seizure are "absences". 80% of the children are reported to have these. My daughter's absences were treated as seizures and had little success in controlling or reducing the number. Of those reported to have these absences 87% give clear indication when they are "back". These findings tie in with "are children with Hydranencephaly conscious". A child cannot "come back" or "out" of something if they aren't conscious in the first place.

Another possible seizure type is the fact that 74% of the children startle easily. 31% of the parents have been told that these are seizures. In many of the children being startled does lead into a seizure.

"Coma" incidents are also fairly common in our children (38%). Again, some parents have been told that these could be seizures. My child's neurologist called them "non convulsive seizures". There doesn't seem to be a clear pattern as to when they occur or how long they last. I know some parents who have sought medical attention for their children when they're in these episodes and have been told that their child is dying or is "shutting down". Granted, a child may be in a coma near the end of their life, but as so many people have reported these occurrences and their child is still living it's likely not a valid

explanation in most of the children. A few children (exact number not known) have managed to be hooked up to an EEG while in this state and it has been shown they were having a non convulsive seizure.

Feeding

Feeding is a common difficulty in children with Hydranencephaly. In 2006 59 (60%) of the children had feeding tubes, and 39 (40%) didn't. The age they got their tubes varies wildly but as the years have progressed the number of children with feeding tubes has increased. But, in spite of that, in the 2006 survey 16 % of the children with tubes still eat orally as well as by tube.

Problems that can cause digestive and feeding issues in children are Gastroesophageal reflux (GERD). This is when food or fluids in the stomach come back up the esophagus and may cause the child to vomit. 58 (59%) of the children are said to have this. Treatments for this condition are medications and surgery. The medications used are too many to list here. When medication doesn't work, the most common surgical treatment is the Nissen Fundoplication. This is when the top of the stomach is made tighter so that food can't come back up. 29 (34%) have had this procedure with 26 (93%) reporting it was a success. Another procedure that is commonly done at the same time is the Pyloroplasty where the bottom of the stomach is made wider so food drains more quickly. Only 5 (7%) of the children are reported to have had that procedure.

Most of the children that eat orally eat smooth purees 27 (47%). 13 (22%) eat liquids only, 14 (24%) eat smooth food with a few chunks (junior food), and 4 (7%) eat regular table food.

Physical Development of a child with Hydranencephaly

Children with Hydranencephaly are all severely physically involved. To help with their physical abilities a large variety of equipment, medications, surgery and other methods have been used. This will give you a summary of what the children in this survey have used/had, etc.

Tone indicates how stiff or floppy a child is. In this survey 47(49%) children were said to have tight (high tone), 5 (5%) were floppy and 44 (46%) both high and floppy. High tone is what usually causes the most problem with the children's limbs and bodies.

Specific physical conditions:

(59) 61% have Clonus which is a tremor in the leg not caused by a seizure.

39 (41%) have a subluxed hip-is partly out of the socket, or may move in and out. 24 (28%) have had one or more of their hips totally out of the socket (dislocated). 41 (43%) have scoliosis, a twist or curve in the spine. 19 (20%) have a Kyphosis which is a forward curve in their spines. This is shown as a hunched back. 30 (32%) have contractures (a tightness in a limb that can't be stretched out).

There are several different surgeries that are done to help either prevent a dislocated hip or to repair one. The first surgery is usually a Tenotomy (a tendon is cut). This is usually done to the tendons at the

top of the legs (adductors-those that allow the legs to go out). It's called a soft tissue surgery and 15 (14%) have had this surgery. If a hip is totally out of socket a bony procedure which uses several osteotomies (cutting the bone). This is basically reconstructing the hip joint and is a very invasive difficult procedure. 9 (8%) of the children have had this procedure done. In some children there are repeated hip dislocations and it's decided to just remove the head of the femur. At the time of this survey 2 (2%) of the children have had this done. It's considered to be an extreme type of surgery so isn't done very often at this time, but it removes any hip pain from the child permanently.

In most children with Hydranencephaly scoliosis is treated with bracing and positioning. But 2 (2%)of the children have had spinal fusion (rods placed). This also is a severe surgery but in some of the children they are healthy enough to tolerate it and have done well since the surgery. Untreated scoliosis can have a very bad effect on both the respiratory and digestive systems. There is no uniform way of preventing scoliosis. You simply do the best you can for your child.

Medical treatment of spasticity

Spasticity is basically stiffness or high tone. It is a major problem in children with Hydranencephaly. Over the last 15 years or so, medications have been developed which can reduce the child's tone and cut down the risk of having contractures.

Baclofen is a medication used to lower tone. It can be given either orally or through an intrathecal pump. This is a reservoir that is placed under the skin. It has a catheter that goes directly into the fluid filled area surrounding the spinal cord which delivers small amounts of medication. This helps reduce spasticity. But, it involves surgery and frequent trips to a medical facility to have the reservoir refilled. 4 children have had this pump inserted. 2 reported it worked the best when compared to oral Baclofen. There has been some mention over the years that Baclofen can cause seizures. This was thought to be true in only 7 (23%) of the children. It is possible that there are fewer seizures when the Baclofen is delivered via pump but there is no data to support or refute that at this time. In the cases where seizures have increased the parents have felt the benefit to outweigh the risk of seizures.

A different treatment for spasticity is injecting substances into muscle groups. These are Botox and Phenol (alcohol) Most commonly used is Botox. 17 of the children have had these injections with 16 saying they worked. Only 4 had phenol injections with 3 saying they worked. I believe the phenol is an older substance that isn't used often now. The problem with Botox is that it has to be injected directly into each muscle group it is wanted to relax. That is painful and can mean multiple injections at one time and the effect only lasts about 3 months. But, it's a good way of preventing surgery or even just seeing if surgery will have the desired effect. As is when it's injected into the adductors (muscles at the top of the legs that when tight can pull hips out of joint). If after the injection the hips are back where they're supposed to be or the position has improved it may tell the dr that surgery to snip the tendons would prevent further problems with hips.

In order to prevent contractures or other orthopedic problems a variety of splints (orthotics) can be used. The first orthotic usually used is called a ankle foot orthotic (AFO). 29 (32%) of the children have AFOs. To prevent contractures in the hands and wrists a hand and thumb splint is worn. In 29 (33%) they

were worn during the day and in 13 (14%) they were worn at night. Another orthotic is called knee immobilizers which are long leg splints. 8 (9%) of the children have used these. A back brace (called a TLSO thoracic Lumbar sacral orthotic) is worn by some children to prevent or control scoliosis. 9 of the children (10%) have used this. A abduction brace can be used to either support children's hips after hip surgery or to prevent hip problems. In the past, the most common way of preventing contractures and orthopedic problems has been through range of motion exercises where a joint is stretched repeatedly and held in a good position for a few seconds of minutes. Now it's thought that sustained positioning over a period of several hours-8 is best can have more long term effects. That's why a lot of children wore their orthotics at night.

In addition to orthotics and braces, equipment is used to help position our children. Another way of stretching or sustained positioning can be the use of a "stander". This is a device that a child is strapped to and then raised to a standing position. 32 (34%)of the children use this equipment. There is also a gait trainer which is a walker for older children. 6 (6%) of the children have used this.

One item almost all children with Hydranencephaly will need at some point in their lives is a wheelchair. In the survey only 63% were said to use a wheelchair. But, almost 25% of the children in this survey are below the age of 1. As the children get older they will almost certainly need a wheelchair. There are a variety of types available with younger children getting a stroller type chair (24) such as a Kid Kart or Convaid Cruiser. Older children will get a standard wheelchair such as a Quickie or Action Tiger (37).

The next most needed piece of equipment is a car seat. As children get older many of the families have adapted vans and tie downs so that their child remains in his/her wheelchair when transported. Apart from that, all children need car seats. Only 73 out of the 106 children represented in this survey were said to use car seats. There are standard car seats that can be used until the child is several years old and special needs car seats for older children.

As the children get older and heavier they will need to be lifted by a mechanical lift. 18 children are using lifts with 10 using a ceiling track lift (either permanent or mobile) and 8 using a portable lift such as a Hoyer.

A child with Hydranencephaly will need a more supportive bed as she/he gets older. Another factor in what type of bed to use is ease of use for the caregiver. Most people find that it works better to have the bed raised higher in order to protect their backs. As many of the children in this survey were under the age of 1, a large number of children are sleeping in standard cribs. 14 sleep in a standard bed, 9 in a manual hospital bed, 17 in an electric hospital bed, 3 in a custom made bed and 6 in a bed called "sleep safe".

Physical abilities of a child with Hydranencephaly

Can your child:	yes	no	Used to	% yes
Roll	28	61	9	29%

Move hands/arms	67	6		68%
Move fingers independently (plucking or pinching)	22	73	2	23%
Move legs	66	6		69%
Kick repetitively or "bicycle"?	53	41		56%
Touch or manipulate objects	22	68	4	23%
Pick up items with his/her hands	5	83	3	5%
Bring hands to midline	41	48		46%
Walk*	0	93		
Sit unassisted	5	88		5%
Sit while supported by legs only	8	80	1	9%
Crawl	4	89	2	4%
Move on the floor other than by crawling	19	66	6	21%
Scoot with wheeled device	5	84	3	5%

* 2 children were reported to walk using a gait trainer (walker)

As you can see, many of the children can move although it's limited. Most can't move much but some can. But, don't count on your child not doing something. My theory was always if we don't try whatever (walking, crawling, etc) then for sure she won't do it.

Head control in the children was mostly poor 48, none 26, good 13 and changes 8. Head control is something that can improve with good positioning or devices such as a back brace to support the trunk. But it's something that is almost always a problem in the children.

Social/Emotional

25 (26%) of the children are said to give hugs and kisses. 89 (91%) of the children cry.

Mood is a common factor among the children. Before the age of 1 26% were said to be happy, 14% quiet and 60% said to be irritable. But, after the age of 1 it changes to 59% happy, 26% quiet and 13% irritable. 2% were answered as "can't tell). This is almost a complete reversal between one year and the next. It is thought that this is because during the first year the brain is settling and trying to work out ways to work despite damage or missing areas. Once the brain stem has realized that it's all there is, so

to speak things calm down and children are happier. So, if you currently have a child under the age of 1 who is crying constantly, take heart, it should get better.

Cognitive Development: this topic will be addressed in the paper titled Cognitive Development and Awareness in a Child with Hydranencephaly which will be released/posted in the near future.

Vision/Hearing:

28 (32%) of the children are reported as seeing. 36 (40%) can see sometimes. We know they aren't going to be able to see as we do, but at least they see some things. An open ended question was asked about what a child sees. Many of the answers were the same with the most common items being light, or lighted objects which was mentioned 25 times, and bright or large colors which was mentioned 13 times. Other things mentioned were distance from object, with many seeing best very close up, black and white, people not objects and objects not people. And many said they weren't sure what their child saw.

A series of questions was asked regarding specific visual responses in children. 68 (77%) of the children react to having room lights turned on or off. 56(66%) of the children turned their heads to or away from visual objects, 39 (51%) turned their eyes to or away from visual objects. 19 (24%) had other reactions to conspicuous visual objects or events. These included reaching towards objects seen, make noises, tune out when overwhelmed with visual stimuli. 11(13%) would peer around an obstacle to look at something. 50 (57%) of the children have nystagmus (flickering of the eyes). 10 (27%) of the children wear glasses. 55 (65%) have been diagnosed with Cortical Visual Impairment which means that their eyes work, it's their brain that's not interpreting what they see. 23 (27%) have been diagnosed with Optic Nerve Hypoplasia. All that means is that their optic nerve isn't properly developed.

87 (94%) of the children can hear. This is in stark contrast to the number of children that can see. This is because a large part of hearing can happen in the brain stem. They won't understand the sounds to as great a degree as do people with a full cortex but they do certainly hear.

81 (86%) turn their head to sound, and 67 (74%)turn their eyes toward sound. 66 (74%) have other reactions to sounds. These include listening carefully, startling, smiling and others. 57 (69%) are most interested in listening to things such as mom's voice, music, bells, stories and much more. Probably every child has some sound they respond to more than anything else. 83(93%) make sounds. Only 13 (14%) say any recognizable words. Again, this corresponds to areas of the brain that may be damaged or missing in a child with Hydranencephaly. 29 (34%) of the children seem to understand specific words. 59 (56%) respond to hearing their own names. 26 (29%) are said to echo what they hear.

14 (16%) use a communication device. Most use the "Big Mack" which is a large colored switch that can have messages spoken into it that play when it's pressed by the child.

Play

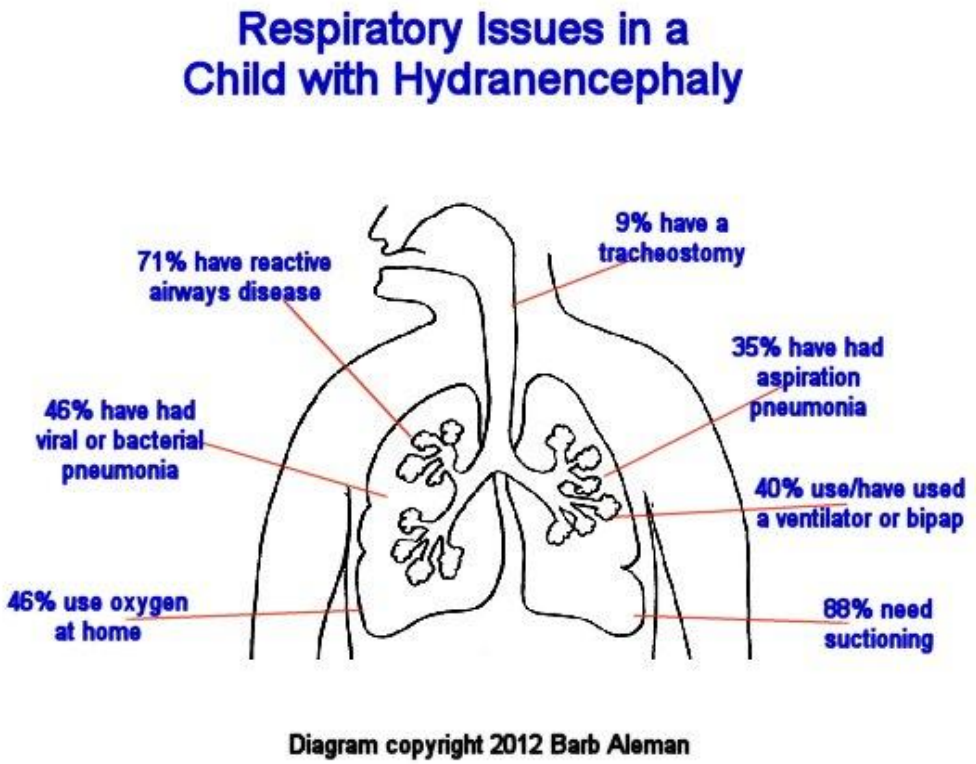
14 (16%) will take turns in play activities. 27 (31%) areable to build up play sequences in which the child gets more and more excited as it goes on. 31 (34%) show signs of wanting or indicating they want more play.

School

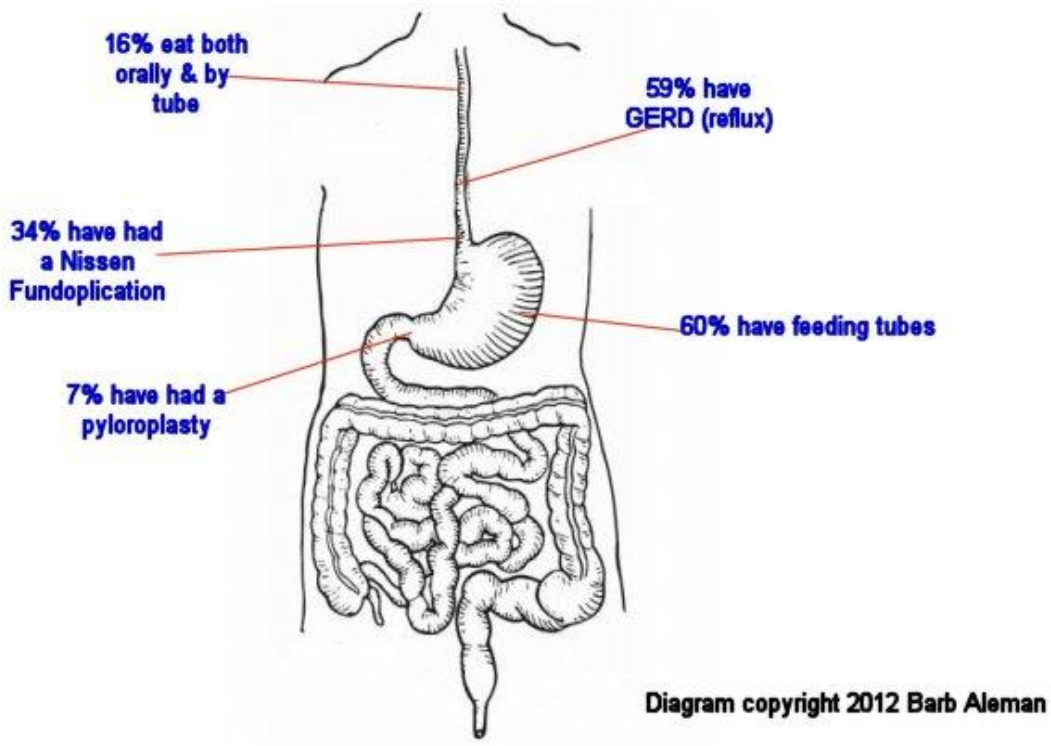
23 (25%) of the children attended school, 16 (18%) didn't, 31 (34%)were too young, 11 (12%) were home schooled and 10 (11%) used to attend school. Of the children who attended school 10(29%) were included with typical peers, 15 (44%) were in a special class, 9 (26%) were both included and in a special class.

Pictorial Summary of the results of this survey

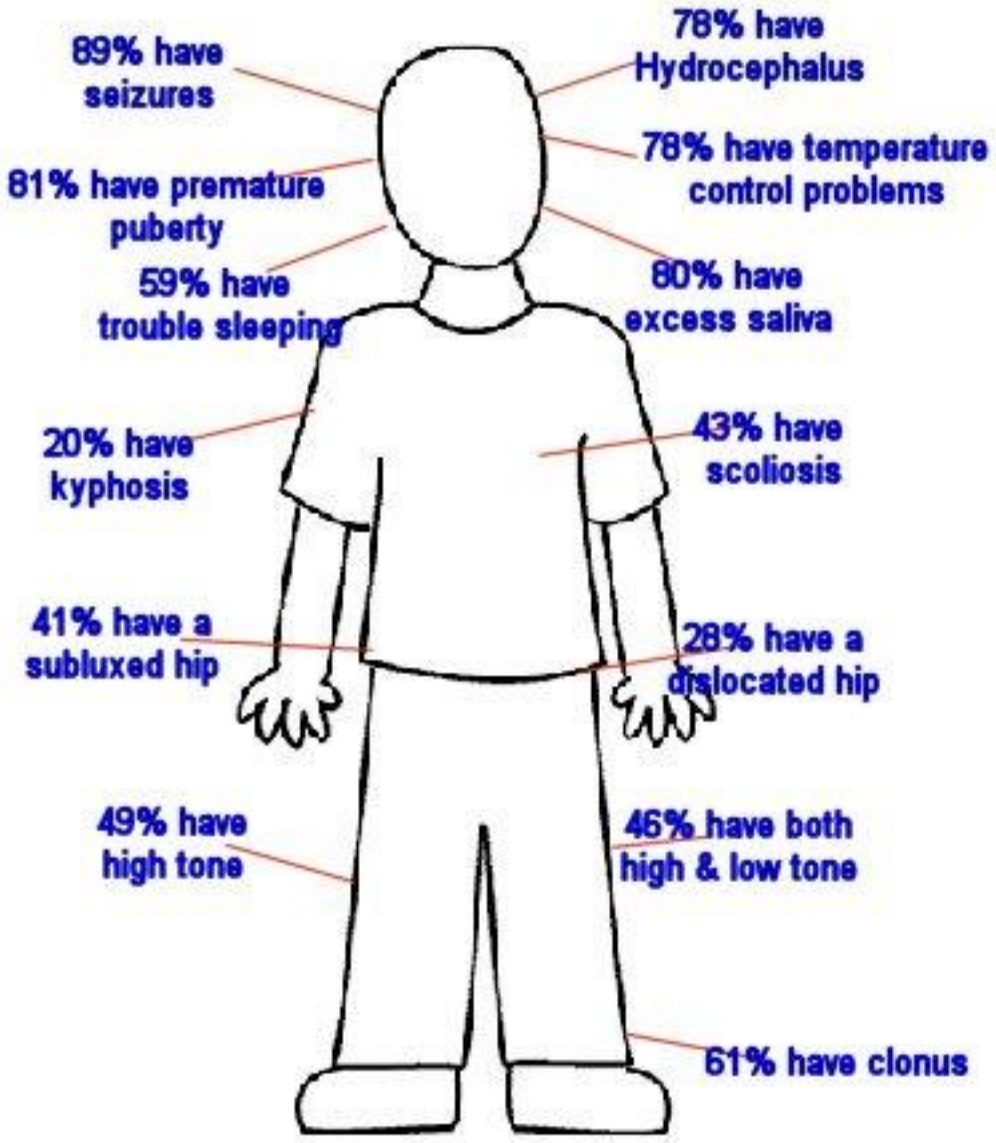
The following are a pictorial summary of the results of the survey. They are meant as an easy reference to the numbers presented in the survey. Remember, though, that this is based only on the results of this survey so aren't necessarily true for Hydranencephaly in general.



Digestive Issues in a child with Hydranencephaly



Health Issues in a child with Hydranencephaly



Physical Capabilities of a Child with Hydranencephaly