Vision/Hearing
In our study, 91% of the children can hear. Some children may not understand what they hear but do respond to sounds and familiar words. However, parents feel that 30% of the children understand at least some of what is said to them. 63% of the children are thought to see at least some of the time. Most of the children have what is called Cortical Visual Impairment. This means that there is nothing wrong with their eyes, but that their brains aren't able to tell them what they are seeing. This sort of vision can come and go so that children see some things and not others or will see one day and not the next.

Other Conditions Children with Hydranencephaly may have:

Asthma/Reactive Airways disease (RAD): 34%.
Diabetes Insipidus (a high sodium level and excessive urine output): 13%
Gastro esophageal Reflux (food comes up which causes vomiting or excessive acid production): 57%
Constipation: 75%

Some children also develop obstructed or difficult breathing and need to have a tracheostomy. At this time 12% of the children have a trach. Several of the children have also needed help from a Bipap or a Ventilator as they get older.

Prognosis: *The outlook for children with Hydranencephaly is poor. Death generally occurs before age 1 (NINDS)*

Of the 350 children we have information on, 162 are living and 188 have died. 16% died before their first birthday. Out of those who have died 31%** died before their first birthday. However of the children who are alive, only 5 are under the age of 1. Our oldest member is 32. It appears that the first year is the most difficult for our children. If they survive that year then many will live a good number of years further.

Raising a child with Hydranencephaly has many challenges but also many joys. You will learn that caring for your child cannot be done alone. You will need the services of a variety of health professionals. But, you will also become the expert on what is best for your child. With love and care a child with Hydranencephaly can have a full active and happy life. They can even attend school with typical children, go on trips, attend parties, and participate in community activities.

Please note: All the "statistics" here only represent what we feel are a small percentage of the actual number of children with this condition around the world. All the families have some sort of computer access to have found us. Therefore these numbers may not be a true representation. The majority of the families live in the US. We're in contact with a few that live in the UK, Australia, New Zealand, Canada and then a few in other countries. . There are also a number of families who chose not to participate in the survey.

*Survey was conducted via a questionnaire online. The 1st had 107 participants and was conducted from 2005-2007 & the 2nd had 106 participants and was conducted from 2010-2011.
** This number includes those children who were still born, those where Drs insisted on a too early delivery, those for whom treatment was denied, and those who only lived a few days due to severity of their condition.

References:
NORD (National Organization of Rare Diseases) website: http://www.rarediseases.org/search/rdbdetail_abstract.html?disname=Hydranencephaly

National Institute Of Neurological Diseases and Stroke website:
http://www.ninds.nih.gov/health_and_medical/disorders/hydranen_doc.htm

Neurology Channel Cephalic Disorders
http://www.neurologychannel.com/cephalicdisorders/types.shtml

Rays of Sunshine
Hydranencephaly Information Network

Hydranencephaly:

The "Facts" according to our families

Information is the Key to Hope

This pamphlet is put out by a group of parents of children with Hydranencephaly and is not meant to take the place of the expertise of a Dr. To learn more about our children and our group please go to:
http://hydranencephaly.com

Hydranencephaly

The "Facts" According to Parents

Disclaimer

Please note: Barb Alemán on behalf of the International Hydranencephaly Support Group writes the following information. We are families of children with Hydranencephaly, not physicians. The information is based on our experiences and on the results of several informal surveys*, which we have done. It is meant to be a balance between what information is currently available and what Doctors tell parents. Text in italics is quotes from a variety of sources on the internet.

Description: *Hydranencephaly is a rare neurological condition in which the brain's cerebral hemispheres are absent and replaced by sacs filled with cerebrospinal fluid*. (National Institute of Neurological Disorders and Strokes NINDS) The amount of brain tissue that each child with Hydranencephaly has, varies from child to child. Many of the children are missing most of their brain tissue above the brain stem. Others have large sections of such tissue left, for example parts of the cortex along the midline or in the occipital lobe (the cortex at the back of the head). *Sometimes the cerebellum and basal ganglia are present and normal. (http:// chorus.rad.mcw.edu/doc/00194.html).* Sometimes they are also missing. Or there are small pockets of brain tissue throughout. Some children also have damage to their brain stem in addition to loss of tissue above the brainstem.

Hydranencephaly is considered to be an extreme form of porencephaly (a rare disorder characterized by a cyst or cavity in the cerebral hemispheres) and may be caused by vascular infections or traumatic disorders after the 12th week of pregnancy. (NINDS)

Cause of Hydranencephaly

In our study, the cause was thought to be Prenatal Stroke: 43%, Prenatal drug exposure: 5%, Prenatal infections: 4%, Death of Twin in Utero: 3%, and in 34% the cause was unknown. In two of the children (1%) the damage occurred after birth. Although physicians don't consider damage after birth to be Hydranencephaly, as their CT scans look the same as the other children, we count them in our numbers.

Children with Hydranencephaly usually appear normal at birth. Sometimes the head is enlarged. *"The infants' head size and spontaneous reflexes such as sucking, swallowing, crying, and moving the arms and legs may all seem normal. However after a few weeks the infant usually becomes irritable and has increased muscle tone. After a few months of life seizures and hydrocephalus (excessive accumulation of cerebrospinal fluid in the brain may develop). (NINDS)*

Treatment: *There is no definitive treatment for Hydranencephaly. Treatment is symptomatic and supportive. Hydrocephalus may be treated by a shunt (a surgically implanted tube that diverts fluid from one pathway to another).*

Hydrocephalus

Hydrocephalus is often one of the first difficulties faced by the child. In our experience 78% of the children have had Hydrocephalus and 75% have needed a shunt. Some children have mild hydrocephalus but live their entire life without a shunt and some never do develop an increase in intracranial pressure from fluid.

Irritability

A child with Hydranencephaly is often very irritable in the first year. A build up of fluid in their heads is one of the first things to check if your child is irritable. As 57% of the children have gastro esophageal reflux, which can be very painful this is something that should also be checked. As the children grow older, 90% are said to be happy or quiet. Only 6% were still described as being fussy or irritable past the one year mark.

Increased Tone

Children with Hydranencephaly may have increased tone in their bodies (spasticity). In our study, 48% of the children were described as having increased tone, and 5% were described as being "floppy". Another 46% were described as having mixed tone. This means that they are sometimes very floppy and sometimes very tight. Physical and Occupational therapy are very helpful in preventing problems from increased tone. However, children do frequently need various orthopedic surgeries as they get older. The therapists will likely suggest a variety of equipment that will also help in preventing a child from developing problems. The children are sometimes referred to as having Cerebral Palsy or Spastic Paralysis.

Seizures

Seizures are fairly common in children with Hydranencephaly (82%). The type and severity of the seizures vary from child to child. Most of the seizures are fairly short and mild but some children do have longer more severe seizures. There are many different seizure medications children take. Often a child needs more than one medication at a time and they frequently need to have their medications changed. A problem both in diagnosing a child who has Hydranencephaly with seizures and in treating them is that EEGs, and medications are geared towards those who are having cortical seizures. Most children with Hydranencephaly have no cortex and have brain stem seizures. So, you and your Dr may agree that your child is having seizures, but nothing shows up on the EEG. And, medications geared at controlling cortical seizures may not work well on a child with Hydranencephaly. That is why your child may need a variety of medications.

Feeding

Children with Hydranencephaly may have trouble sucking and swallowing. Many families are told that their child will lose these abilities within a few weeks of birth when the "higher" cortical areas of the brain normally kick in. We haven't found this to be true. In our study 33% of the children eat orally, 46% eat only by tube and 19% eat both orally and by tube. As the child gets older he may have more trouble eating and eventually need a feeding tube.