What is Hydranencephaly?

In a fetus/baby with hydranencephaly, a severe anomaly in the development of the brain has occurred. The skull is filled with a sac-like structure containing fluid and replacing the superior brain structures, with only some of the lower brain structures still present. This is a very severe condition associated with the death of the baby before or after birth and severe handicaps if the baby survives.

How does hydranencephaly happen?

The most accepted theory is that the occlusion of vessels supplying the higher and anterior parts of the brain has caused its destruction.

How are chromosomes relevant to hydranencephaly?

Hydranencephaly can sometimes be seen in babies with an abnormal number of chromosomes, but in the majority of hydranencephaly cases, there is a normal number of chromosomes. A rare variant of hydranencephaly occurs in a syndrome called Fowler that can run in families.

Should I have more tests done?

Hydranencephaly is usually suspected after ultrasound examination of the baby’s head which appears “empty” and filled with cerebral fluid. The diagnosis usually happens at the mid trimester scan, but it can also be seen at earlier scans. The doctor may suggest doing an amniocentesis (taking some fluid from around the baby) to check the chromosomes.

What are the things to watch for during the pregnancy?

The pregnancy usually progresses normally, and you will feel normal baby movements. In some cases, the baby’s head grows too little (microcephaly) or too much (macrocephaly), due to accumulation of cerebral fluid in the skull. For this reason, the doctor may monitor the growth of the baby’s head with more scans. In addition, the size of the baby’s head, particularly if it is larger than usual, may influence the route of delivery and care of the baby during the delivery process.
**Hydranencephaly**
*Patient Information Series – What you should know, what you should ask.*

**What does it mean for my baby after it is born?**

It is advisable that your baby is born in a center with experience in managing these rare anomalies. There is no cure for hydranencephaly and the only cure that can be offered is to ease the symptoms of the baby that may have seizures or breathing difficulties. A drain may also be placed in the head to stop the fluid to accumulate in the head. Babies with hydranencephaly are severely handicapped and usually have a short life expectancy. This is the reason why your doctor will discuss with you the option of termination of pregnancy, if this is an option.

**Will it happen again?**

Hydranencephaly is usually an isolated accident that will not happen again.

**What other questions should I ask?**

- Does this look like a severe case of hydranencephaly?
- Is it possible for me to terminate the pregnancy?
- Where should I deliver?
- Where will the baby receive the best care after it is born?

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