Hydrocephalus

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Definition

Hydrocephalus relates to the progressive enlargement of the ventricular structures in the brain.

Introduction

Within the brain there is solid tissue and hollow cavities. The solid portions are called hemispheres and the hollow areas or cavities are named ventricles. These "cavities" in the brain play very important roles. It is in this section of the brain that the cerebrospinal fluid (CSF) is produced and absorbed. The CSF is important in lubricating and protecting the brain; it is also believed to carry nutrients through out the brain and spinal canal. The CSF circulates around the brain and the spinal cord. There are common pathways through which the fluid is produced, goes around and then returns to be re-absorbed. There is a balance between the production and the re-absorption of the CSF that maintains a fairly constant volume.There are however, a number of situations and conditions that can alter this balance causing enlargement of the ventricles with potentially devastating consequences. The accumulation of the CSF with subsequent enlargement of the ventricles is what is called hydrocephalus.

There are many reasons why this can happen. Some of these reasons take place before the baby is born (congenital abnormalities), other times is related to trauma or infection and tumors at any time after birth and through out life.

When babies develop hydrocephalus soon after birth (neonatal) the more frequent reasons are:

- Aqueductal Stenosis 33%
- Myelomeningocele (spina bifida)with
- Arnold-Chiari malformation 28%
- Communicating hydrocephalus 22%
- Dandy-Walker malformation 7%
- Other 10%

When babies have the hydrocephalus in uterus (fetal) it is usually more severe and it is associated with anomalies of the brain in 60-65% of the cases.

After the neonatal period other causes of hydrocephalus are more common such as complications from:

- Meningitis
- Intracranial bleeding (hemorrhage)
- Tumors
- Calcifications
- Cysts
- Secondary to toxins

Identifying the original cause of the hydrocephalus is important because of the treatment and prognosis implications.

Diagnosis

Although there are physical signs and symptoms suggestive of hydrocephalus this condition needs confirmation by CT or MRI of the brain. In the newborn ultrasound can be very helpful too. The clinical manifestations of this

condition depend on the age of the individual. Symptoms are due to increased intracranial pressure due to accumulation of the CSF beyond what it is physiologic.

In the very young: the head size enlarges, the fontanel (soft spot) may be bulging, there may be feeding problems because of depressed neurological status and other symptoms or findings related to the cause of the hydrocephalus.

Once the soft spot closes and the skull bones are fused together, the signs of increased intracranial pressure become more severe. There is frequently:

- Nausea & vomiting
- Headache
- Visual problems (like double vision)
- Lethargy
- Ataxia (loss of balance and coordination)
- Seizures

Sometimes even coma and death.

Some of these symptoms may develop gradually or may be sudden like in cases where there is intracranial bleeding.

There are occasions in which the ventricles are enlarged but this is not due to obstruction of CSF or problems with absorption of the same. In situations where the hemispheres did not develop normally or due to scarring they "shrink", the ventricles then enlarge to compensate for the volume loss. In these cases there is no increased intracranial pressure and characteristically there is no need for drainage or a shunt mechanism. This is sometimes called normal pressure hydrocephalus.

Prevention and Treatment

Preventive measures can be divided into those directed at prevention of fetal anomalies and those interventions related to preventing head trauma, infections like meningitis and anticipating complications from intracranial masses.

A very successful intervention to prevent neonatal hydrocephalus is increasing the oral intake of Folic Acid by women of child bearing age. This intervention reduces spina bifida (Myelomeningocele). Prevention of congenital infections like Toxoplasmosis and Cytomegalovirus can also decrease cases of hydrocephalus.

The increased use of helmets in sports and seat-belts are also excellent preventive measures to decrease trauma to the brain.

Treatment strategies once the diagnosis is made need to address the damaging effects of increased intracranial pressure. This usually means surgical intervention. The accumulation of CSF with progressive enlargement of the ventricular structures requires an alternative system (shunts) to drain the excess of CSF and therefore decrease the intracranial pressure. Before there were shunts available the mortality was very high; the availability of silicone shunts have decreased mortality to 15% (Hirsh, et al). Neurosurgeons are the specialist that place these shunts and evaluate for malfunctions.

There have been many shunt mechanisms designed over the years. However, the most commonly used connect the ventricles to the abdominal cavity and are referred to as VP Shunts. Other shunts can be placed from the brain into the heart, the pleural space or from the spinal canal to the abdomen. These shunts have several components; there is a portion that goes into the ventricles, a pump or valve mechanism and a longer draining tube. Some have a reservoir compartment but not all of them. It is important to know the components because of the potential for complications on the different sections. Again, neurosurgeons are the specialists that need to evaluate the proper function of these

shunts. Shunt evaluation needs to happen at the hospital where immediate action can be taken, if necessary in the operating room.

Emergency Situations – What can go wrong?

If a hydrocephalus is not detected within reasonable time the effects of the increased intracranial pressure can be devastating. The increased pressure to vital brain structures can lead to arrhythmias, irregular breathing, hypertension, severe vomiting, headache and possibly death. Cranial nerves can also be affected causing blurred or double vision. In the very young child this increased pressure can also have a deleterious effect in normal brain growth causing a decrease in intellectual acuity.

There are well known complications once the shunt is in place. Infections, dislodgement, plugging of the shunt can take place at any time/age. The rate of complications from shunt malfunction decrease with time. Most complications happen soon after placement.

Conclusion

Hydrocephalus is a non-physiologic accumulation of CSF in the brain causing enlargement of the ventricles. It is commonly associated with central nervous system malformations, trauma, infections and or intracranial masses that result in increased intracranial pressure. The complications can be very serious and long lasting. Emergency surgical intervention is often necessary.

References

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