Porencephaly

Porencephaly is a rare disorder of the central nervous system. It involves cysts or cavities in a cerebral hemisphere. The cysts or cavities are usually the remnants of destructive lesions. These are due to stroke or infection. But they are sometimes the result of abnormal development. The disorder can occur before or after birth. Most infants show symptoms shortly after birth. Diagnosis is usually made before age 1.

SYMPTOMS

Signs may include:

- Delayed growth and development.
- Slight or incomplete paralysis (spastic *hemiplegia*).
- Low muscle tone (*hypotonia*).
- Seizures. These are often infantile spasms.
- Large head (*macrocephaly*).
- Small head (*microcephaly*).

Individuals with this disorder may have:

- Poor or absent speech development.
- Epilepsy.
- Hydrocephalus.
- Spastic contractures. This is shrinkage or shortening of a muscle.
- Mental retardation.

TREATMENT

Treatment may include:

- Physical therapy.
- Medication for seizure disorders.
- A shunt for hydrocephalus.

The prognosis for individuals with this disorder varies according to the location and extent of the lesions. Some patients with this disorder may develop only minor neurological problems and have normal intelligence. Others may be severely disabled. Others may die before the second decade of life.

RESEARCH BEING DONE

The NINDS conducts and supports a wide range of studies. These explore the complex mechanisms of normal brain development. The knowledge gained from these studies provides the foundation for understanding how this process can go awry. It offers hope for new means to treat and prevent developmental brain disorders, including porencephaly.

Research is continually being done to learn more about this problem.

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