

Hydrocephalus

(“Water on the Brain”)

Basics

OVERVIEW

- The cerebrum is the large rounded structure of the brain; it is composed of two hemispheres; inside each hemisphere is a cavity, known as a “lateral ventricle”; the two lateral ventricles and two other ventricles (third and fourth ventricles) form the “ventricular system” of the brain; cerebrospinal fluid is produced from specialized areas in the ventricles; the cerebrospinal fluid flows between the ventricles and into the spinal canal; blockage of flow can lead to increased pressure in the lateral ventricles, resulting in hydrocephalus
- Cerebrospinal fluid (also known as CSF) is a specialized body fluid that cushions the brain and spinal cord
- Hydrocephalus is a condition in which the ventricular system of the brain is abnormally dilated due to an increased volume of cerebrospinal fluid
- May be symmetrical or asymmetrical
- May involve the entire ventricular system or only parts of the ventricular system if a blockage of the flow of cerebrospinal fluid is present, in which case, the CSF volume increases in the part of the ventricular system behind the blockage (in other words, the blockage acts as a dam to the flow of CSF)
- Lay term for hydrocephalus is “water on the brain”



GENETICS

- Inherited hydrocephalus—Siamese cats and Yorkshire terrier
- Siamese cats—autosomal recessive

SIGNALMENT/DESCRIPTION OF PET

Species

- Dogs
- Cats

Breed Predilections

- Congenital (present at birth) hydrocephalus—small and short-nosed, flat-faced (known as “brachycephalic”) dogs: bulldogs, Chihuahuas, Maltese, Pomeranians, toy poodles, Yorkshire terriers, Lhasa apsos, cairn terriers, Boston terriers, pugs, and Pekingese
- Inherited hydrocephalus—Siamese cats and Yorkshire terriers; high incidence of enlargement of the ventricles (known as “ventriculomegaly”) that do not cause clinical signs in normal adult beagles
- Acquired (condition that develops sometime later in life/after birth) hydrocephalus—any breed of dog or cat

Mean Age and Range

- Congenital (present at birth) hydrocephalus—usually becomes apparent at a few weeks up to 1 year of age; sudden (acute) onset of signs can occur in dogs with previously undiagnosed congenital hydrocephalus—the exact cause of this sudden onset of signs is uncertain
- Acquired (condition that develops sometime later in life/after birth) hydrocephalus—any age

Predominant Sex

- None

SIGNS/OBSERVED CHANGES IN THE PET

- Highly variable
- Signs may be sudden (acute) or gradual
- Signs may not change (known as “static signs”) or may progress (known as “progressive signs”)
- Congenital (present at birth) hydrocephalus—may occur without clinical signs, especially in dogs of toy breeds; other malformations or abnormalities of the central nervous system may be present, which may contribute to clinical signs
- Acquired (condition that develops sometime later in life/after birth) hydrocephalus—signs from the underlying disease (that is, the disease that led to the development of hydrocephalus) may be as or more prominent than signs from hydrocephalus itself
- Behavioral signs—decreased awareness; lack of or loss of training ability (including house-training); excessive sleepiness; vocalization; sometimes hyperexcitability
- Visual abnormalities, including blindness
- Seizures—may be noted
- Head—may appear large and dome-shaped; persistent “soft spot” of the head
- Outward and downward deviation of both eyes (known as “bilaterally divergent strabismus”) is present in some dogs with severe congenital (present at birth) hydrocephalus
- Cerebral disease—abnormal behavior, dullness, sleepiness, circling
- Gait abnormalities—incoordination, wobbly or “drunken” gait (known as “ataxia”)
- Severely increased pressure within the head (known as “intracranial pressure”)—stupor or coma, pinpoint or dilated fixed pupils, and abnormal breathing patterns; may lead to fatal condition in which the brain has pushed downward in the skull and has herniated through the opening that leads to the neck (known as “brain herniation” or “tentorial herniation”)
- Severity of clinical signs may not correlate with the size of the ventricles in the brain, although worse clinical signs tend to be seen with greater enlargement of the ventricles

CAUSES

- Congenital (present at birth) hydrocephalus—numerous congenital malformations cause blockage or obstruction of the ventricular system and result in hydrocephalus; inherited malformation of the ventricular system; prenatal infection (dogs—parainfluenza virus; cats—coronavirus); exposure to compounds that cause abnormal development of the embryo (known as “teratogenic compounds”); bleeding into the brain of the fetus or newborn secondary to a difficult birth (known as “dystocia”); nutritional deficiency (vitamin A)
- Acquired (condition that develops sometime later in life/after birth) hydrocephalus—tumors or cancer, abscesses, and inflammatory diseases

RISK FACTORS

- Pets with hydrocephalus that do not have clinical signs may develop clinical signs in the face of an insult, such as infection or trauma

Treatment

HEALTH CARE

- Inpatient—intensive care for pets with severe signs or when undergoing surgical therapy
- Outpatient—pets with mild to moderate signs that can be treated medically
- Prevent secondary complications of lying down for prolonged periods in stuporous or comatose pets—avoid pressure sores; drying eyes; and lung congestion

ACTIVITY

- Depends on severity of signs

SURGERY

- Surgical procedure in which cerebrospinal fluid is shunted from the ventricles of the brain to the abdominal cavity or to the right atrium of the heart—definitive treatment; clinical signs may not resolve completely; residual signs usually indicate irreversible brain damage
- Surgery should be considered only when medical management is ineffective or results in adverse side effects
- Surgery or radiation therapy for a brain tumor or other mass lesion—consider if it is the underlying cause of hydrocephalus

Medications

Medications presented in this section are intended to provide general information about possible treatment. The treatment for a particular condition may evolve as medical advances are made; therefore, the medications should not be considered as all inclusive

- Reduce production of cerebrospinal fluid (CSF)—steroids (such as prednisone or dexamethasone); or carbonic anhydrase inhibitors (such as acetazolamide); omeprazole has been reported to reduce CSF production in the dog in an experimental model, but no data is available on the usefulness of this drug in treating hydrocephalus
- Reduce increased pressure within the head (intracranial pressure)—medications to decrease excess fluid in the body (known as “diuretics,” such as mannitol and/or furosemide); these are short-term treatments only, helpful for immediate treatment of severe cases
- Treat underlying cause—administer specific drugs when possible (such as antibiotics for bacterial infection)

Follow-Up Care

PATIENT MONITORING

- Monitor for increase in severity of signs of hydrocephalus and for signs from an underlying cause (such as a tumor in the skull)

POSSIBLE COMPLICATIONS

- Infection and shunt blockage (in up to 50% of patients) can occur following surgical shunting of cerebrospinal fluid; shunt revision commonly is needed
- Excessive shunting of cerebrospinal fluid may result in severe and potentially fatal complications
- Brain pushes downward in the skull and herniates through the opening that leads to the neck (brain or tentorial herniation) and death

EXPECTED COURSE AND PROGNOSIS

- Good to poor: depends on cause and severity of signs of hydrocephalus
- Mild congenital (present at birth) hydrocephalus—good prognosis; may require only occasional medical treatment

Key Points

- Observe for deterioration in mental alertness, vision, and behavior, which may signal worsening of hydrocephalus
- Severity of the clinical signs may not correspond to the degree of ventricular enlargement of the brain