Hydrocephalus is a condition of enlargement of the ventricular system that can result in the loss of overlying brain structure(s). This ventriculomegaly increases cerebrospinal fluid (CSF) volume with or without associated changes in CSF pressure. High-pressure hydrocephalus (HPH) is due to obstruction of CSF flow either within the ventricular system or at the level of the arachnoid villi within the cerebral venous sinuses. The hydrocephalus may persist beyond a transient, noncommunicating obstructive cause. Normal-pressure hydrocephalus (NPH) is the result of CSF accumulation in the cranial vault no longer occupied by brain tissue with patency of the ventricular outflow pathway. Both HPH and NPH can occur either as congenital or acquired conditions.

The goal in the clinical approach to hydrocephalus is early diagnosis, accurate identification of underlying etiology, and institution of therapy early in the course of disease. The longer the duration of HPH, the greater the irreversible damage to the overlying cerebral structures due to pressure necrosis and chance for brain herniation. Advanced imaging modalities are now available for an accurate diagnosis to allow specific therapeutic approaches to be considered. The correlation between ventricular size and clinical signs, however, is poor in dogs. A more accurate method of diagnosis is direct measurement of intracranial pressure (ICP) and resistance to CSF outflow. These methods, however, are still not routinely used in veterinary medicine. Therefore clinicians must be careful to avoid overinterpreting ventriculomegaly and must direct therapeutic strategies at alleviating the clinical signs.

**DIAGNOSTIC CRITERIA**

**Historical Information**

**Gender Predisposition:** None.

**Age of Onset:** Defined by the underlying etiology. Many animals with congenital malformation of the ventricular system develop clinical signs within the first few months of life. Animals with acquired causes (e.g., inflammation, neoplasia) do not exhibit signs until the onset of the inciting disease.

**Breed Predisposition:** Dog breeds predisposed to congenital communicating NPH are the Maltese, Boston terrier, Chihuahua, English bulldog, Lhasa apso, Pekingese, Pomeranian, Shih Tzu, toy poodle, Yorkshire terrier, pug, and cairn terrier. These dogs are suspected of having communicating NPH, as brain imaging and pressure measurements are often not available. Hereditary hydrocephalus has been reported in the Siamese cat as an autosomal recessive trait and may occur sporadically in other cat breeds.

**Owner Observations**

- **Congenital communicating NPH:** Signs are typically gradual, constant, and often progressive. They are centered around behavioral disturbances, such as failure to be housebroken, learn commands, and socialize. Many of these animals have a smaller stature and quieter demeanor compared with littermates.

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• Acquired HPH: Signs are more acute in onset, progress rapidly, and are characterized by a pattern of dementing behavior (e.g., altered personality, head pressing into corners, failure to recognize or respond normally to owners), “stiff” neck, and abnormal body postures due to brain herniation.

• Seizures may occur with any type of hydrocephalus.

**Physical Examination Findings**

Partial or generalized seizures may occur with any type of hydrocephalus.

The precise characterization of NPH versus HPH requires measurement of CSF pressure (see Other Diagnostic Tests). Therefore the following categories are often based on clinical suspicion of the type of hydrocephalus present.

**Congenital NPH**

(signs from birth; Figure 1): Diffuse forebrain signs are usually more chronic and progressive. Any or all of the following may occur:

- Enlarged cranium with a domed appearance.
- “Open” cranial fontanelle(s).

**Acquired HPH**

(signs develop later in life): Diffuse forebrain signs are often more acute and rapidly progressive. Any or all of the following may occur:

- Normal appearing cranium.
- Normal body size.
- Dementia.
- Visual deficits.
- Cervical pain.
- Head shaking.
- Postural deficits.
- Paresis.
- Papilledema.

Bradycardia and systemic hypertension can be associated with increased ICP.

**Acquired NPH**

(signs develop later in life): Signs are initially due to the loss of brain tissue, either from acute (e.g.,...
infarct) or chronic (e.g., brain atrophy) causes. Also known as hydrocephalus ex vacuo. Acute exacerbation of signs can occur as a result of intraventricular hemorrhage and shearing of the white matter tracts from the lateral ventricle with progressive ventriculomegaly. Any or all of the following may occur:

- Normal appearing cranium.
- Normal body size.
- Dementia.
- Visual deficits.
- Cervical pain.
- Head shaking.
- Postural deficits.
- Paresis.
- Papilledema.

Bradycardia and systemic hypertension can be associated with increased ICP.

**Laboratory Findings**

- Clinical laboratory testing is usually normal.
- Multiple congenital defects may be present, including portosystemic shunting. Serum bile acid study will be abnormal if a portosystemic shunt is present.
- Sodium electrolyte imbalance may develop if a change in thirst occurs due to hypothalamic dysfunction.
  - Hypernatremia can be seen in patients with hypodipsia or central diabetes insipidus associated with hydrocephalus.
  - Hyponatremia can occur with psychogenic polydipsia-associated behavioral changes.

**Other Diagnostic Tests**

- Electroencephalography $—$ A pattern of generalized high voltage (>50 µV), slow wave (<8 Hz) activity is indicative of hydrocephalus. Sensitive but nonspecific diagnostic test that evaluates forebrain function.
- CSF analysis $—$ Cytology, protein analysis, and possibly infectious disease titers are indicated for inflammatory diseases associated with obstructive hydrocephalus (e.g., FIP). Otherwise, CSF analysis is usually not indicated for congenital communicating NPH. Risk of brain herniation exists in cases of obstructive HPH.
- CSF pressure monitoring $$$--$$$ — CSF opening pressure can be measured either from the cerebellomedullary cistern at the time of sample collection or during a ventriculostomy procedure (discussed below). Normal opening pressure from the cerebellomedullary cistern in dogs under barbiturate anesthesia can be obtained from a fluid manometer measurement and estimated using the following equation (Simpson and Reed, 1987):

\[
\text{Opening pressure (mm CSF)} = 46.5 + 3.83 \times BW(\text{kg}) - 0.048 \times BW(\text{kg})^2
\]

Ventriculostomy opening pressure should measure <25 mmHg.

**KEY TO COSTS**

$ indicates relative costs of different diagnostic and treatment regimens.
- $ costs under $250
- $$ costs between $250–$500
- $$$ costs between $500–$1000
- $$$$ costs over $1000

**STANDARDS OF CARE: EMERGENCY AND CRITICAL CARE MEDICINE**

**FIGURE 2**

Transfontanellar cranial ultrasonogram demonstrating suspected communicating NPH of a congenital nature (same dog as in Figure 1).

- ICP monitoring $$$--$$$—Most useful for monitoring therapeutic success of attempts to reduce CSF pressure and volume. Specialized equipment (e.g., Codman® ICP Express), animal sedation, and an invasive approach for transducer placement are required. Normal ICP pressure measured from the parenchymal or subdural space is <25 mmHg in dogs and cats.
- Skull radiographs $—$ Ground glass appearance of skull with open suture lines and fontanelles, thinning of the cortical bone and wing of the sphenoid bone, and loss of the osseous tentorium may be present. Insensitive and nonspecific diagnostic test.
- Cranial ultrasound $—$ Transfontanellar sonographic (real-time B mode using a 5.0–7.5 MHz transducer) evaluation of the ventricular system is noninvasive, rapid, easy to obtain, and reliable, provided an open fontanelle is present (Figure 2). A lateral ventricular height (obtained from a transverse scan) >0.3 cm is considered abnormal for most small breeds, although normal breed-specific dimensions are unknown. “Closed”
CHECKPOINTS

- Glucocorticoid Therapy—The long-term benefit of glucocorticoid therapy in the management of hydrocephalus is not well documented. The ability to confirm the etiology, type of hydrocephalus, and response patterns is necessary to determine the benefit of this therapy in an evidence-based fashion.
- Ventriculoperitoneal (V-P) Shunting—While V-P shunting has been employed in veterinary medicine for several decades, the appropriate outcome measures to determine the longer term risks and benefits of the procedure have been poorly documented in the literature. In particular, if the long-term complication rate parallels that of human medicine, it may be as high as 50%.
- ICP Monitoring—This diagnostic and prognostic methodology is standard in the treatment of hydrocephalus in humans. More information is needed about its applicability in emergency care and surgical facilities.

fontanellar examinations may be possible if the cortical bone is thin. When used for young toy breeds, this can be a sensitive but less specific diagnostic test for hydrocephalus.

- Cranial computed tomography ($$)—CT scanning can provide noninvasive three-dimensional visualization of ventricular size and the entire ventricular outflow tract to detect obstruction. Cost and the need to sedate younger animals are the major drawbacks. While the procedure is sensitive and specific for a diagnosis, correlation between ventricle size and clinical signs may be poor.
- Cranial magnetic resonance imaging ($$$--$$$$) (Figure 3)—MRI of the brain is optimal to evaluate the parenchymal changes associated with hydrocephalus, presence of brain edema, exact location of an obstructive cause, and presence of other abnormalities. Periventricular, interstitial cerebral edema (high T2 signal) is indicative of HPH but may also be seen with inflammatory diseases (e.g., FIP) and neoplasia. High sensitivity and specific, MRI scanning is the preferred test for the identification of obstructive causes of HPH.

Summary of Diagnostic Criteria

- Chronic or acute onset of diffuse forebrain signs, including abnormal behavior, change in vision, gait abnormalities, and/or seizures, may be seen.
- Diagnostic imaging findings consistent with hydrocephalus.
- An abnormally shaped cranium with or without “open” fontanelles is often seen with congenital hydrocephalus.
- Acute onset of forebrain signs is often seen with obstructive HPH.

Differential Diagnosis

Differential diagnoses for hydrocephalus are based on the neurolocalization of diffuse forebrain signs. This list is divided into hydrocephalic and nonhydrocephalic etiologies.

Hydrocephalic Conditions

- Congenital Hydrocephalus
  - Communicating NPH—Hydrocephalus ex vacuo: Accumulation of CSF in space previously occupied by brain tissue. Examples include porencephaly (cystic space), hydrancephaly (lack of cerebral hemisphere formation), and lissencephaly (failure of formation of normal cortical gyri and sulci).
  - Noncommunicating HPH leading to NPH later in life—Hypothesized malformation of the arachnoid villi and/or mesencephalic aqueduct leading to failure of normal CSF outflow in utero or in the early postnatal period. This is the most commonly diagnosed form of hydrocephalus in the dog.

Acquired Hydrocephalus

- Communicating NPH—Hydrocephalus ex vacuo—Accumulation of CSF in space previously occupied by brain tissue (e.g., postischemic event, cortical degeneration from aging).
- Adult onset, normotensive, progressive hydrocephalus of a suspected congenital nature is seen in the dog. The correlation to clinical signs, which may be limited to epileptic seizures and behavior disturbances, is not clear.
- Noncommunicating HPH
  - Cats—Malformation with stricture of the mesencephalic aqueduct, FIP, toxoplasmosis, other causes of ependymitis, intra- or extraventricular neoplasia (meningioma, ependymoma, choroid plexus papilloma), and trauma.
  - Dogs—Malformation with stricture of the mesencephalic aqueduct, encephalitis and/or ependymitis (e.g., parainfluenza encephalitis, parasitic granuloma, Cryptococcus), intra- or extraventricular neoplasia (meningioma, ependy-
moma, choroid plexus papiloma), and trauma.

Nonhydrocephalic Conditions

Metabolic Diseases

- Hepatic encephalopathy (portosystemic shunting).
- Uremic encephalopathy (acute or chronic renal failure).
- Electrolyte imbalances (e.g., sodium disorders).
- Endocrine diseases (e.g., cerebral myxedema from hypothyroidism, ketogenic diabetes mellitus).
- Hypoglycemia.
- Hypoxia/ischemia.

Toxicity/Nutritional

- Lead.
- Thiamine deficiency.
- Sedative/hypnotic drug reactions.

Primary Brain Disorders

- Neuronal storage diseases.
- Primary brain neoplasm with associated cerebral edema.
- Inflammatory diseases (e.g., canine distemper virus, fungal, protozoal, or immune-mediated encephalitides).
- Idiopathic epilepsy.

TREATMENT RECOMMENDATIONS

Initial

Suspected NPH (acquired or congenital etiology)

- Glucocorticosteroids to decrease CSF production.

$-$

- Prednisone: 0.25–0.5 mg/kg PO bid for 7–14 days initially, then taper to an acceptable daily to every other day dose (around 0.1 mg/kg/day).

- Diuretics to decrease CSF volume.

- Furosemide: 1 mg/kg PO q12–24h or

- Acetazolamide: 10 mg/kg PO q8h (dogs only).

Suspected HPH (acquired or congenital etiology)

- Glucocorticosteroids to decrease CSF production.

- Prednisolone sodium succinate: 10 mg/kg IV q6h for 24 hr or

- Methylprednisolone: 15–30 mg/kg IV initially, then 5 mg/kg q8h for 24 hr.

- Follow up with prednisone therapy (0.25–0.5 mg/kg PO bid for 7–14 days initially, then taper to an acceptable daily to every other day dose [around 0.1 mg/kg/day]).

- Mannitol (20%)—Hypermolar agent to rapidly reduce ICP due to interstitial cerebral edema formation.

- 1 g/kg IV over 20 min. An additional two doses (0.25–0.5 g/kg) may be given within 24 hr.

- Restricting fluids to approximately one third of urine output for the next hour will reduce the effect of “rebound” intracranial hypertension.

- Furosemide—Prolongs the ICP-lowering effect of mannitol by decreasing cerebral blood volume and CSF production.

- 0.7–1.0 mg/kg IV once at termination of the mannitol therapy. May be repeated twice in 24 hr.

- Hyperventilation—Assisted hyperventilation under general anesthesia can rapidly lower ICP by inducing vasocostriction (useful when other medical management is insufficient).

- Requires general anesthesia, endotracheal intubation, and monitoring of end tidal PCO₂ and arterial PaCO₂.

- Maximal effect occurs when the PaCO₂ is 25–30 mmHg. Excessive hyperventilation may lead to cerebral ischemia due to excessive vasocostriction.

- Gradual return to normoventilation is required to prevent rebound intracranial hypertension.

- Barbiturate therapy—Barbiturates decrease cerebral metabolism and cerebral blood flow and thus decrease ICP.

- Propofol (a short-acting barbiturate that can be titrated): Initial dose of 5–10 mg/kg IV and titration to effect with CRI can be used in conjunction with hyperventilation.

- Antiepileptic drug therapy for treatment of seizures.

- Diazepam (emergency treatment)—0.5 mg/kg IV or 1–2 mg/kg per rectum.

- Phenobarbital (maintenance therapy)—2.5 mg/kg PO bid.

- Potassium bromide—40 mg/kg/day PO with food; can be divided bid.

Alternative/Optional Treatments/Therapy

Ventriculostomy Placement $$

Indication

Acute, emergent HPH with associated or impending signs of brain
herniation and failure of medical therapy as described above.

**Comments**
- Requires general anesthesia with appropriate monitoring.
- CT is recommended prior to the procedure if feasible to locate possible obstructive cause.

**Procedure**
- A rostralateral approach to the lateral ventricle is made, approximately 2 cm lateral to the midline, at the level of the bregmatic cranial suture.
- A dorsoventral skin incision is made, followed by dissection of underlying temporalis muscle with retraction to the bone. The bone is scraped with a periosteal elevator to prevent penetration of soft tissue into the brain.
- A burr hole is made with a handheld or pneumatic drill using a 3 mm rounded drill bit. A sharp pointed drill bit can be used with a handheld drill and guide, provided an appropriate depth gauge stop is used to prevent excessive penetration of the bit.
- Slowly insert ventriculostomy catheter (Elserg ventricular catheter, 4-in., 5–7 Fr; Codman®, Johnson & Johnson).
- Remove stylet at approximately 0.5 mm intervals to check for CSF flow.
- Drain CSF until normal pressure range (opening pressure, 25 mmHg) is detected or flow of CSF slows significantly. Pressure can be monitored with a pressure transducer or a fluid manometer.
- Submit CSF for cytologic analysis and culture.

**Ventriculoperitoneal (V-P) Shunting**

**Indications**
To reestablish brain function by normalizing ventricular pressure and cerebral blood flow.

**Comments**
- Requires general anesthesia with appropriate monitoring.
- CT or MRI is necessary prior to the procedure to evaluate the presence and location of an obstructive cause and degree of hydrocephalus.
- CSF analysis is necessary when an infectious cause is suspected.
- The Codman® Hakim Precision Valve medium- to low-pressure range (70 ± 10 mmH2O) ventriculoperitoneal shunt system is recommended. An appropriate length tube is used to reach from the ventricle to the peritoneum with sufficient excess for movement and/or growth.

**Procedure**
- A ventricular catheter is placed via a burr hole and durotomy into the lateral ventricle and secured.
- The one-way valve system is primed, tunneled subcutaneously from the caudal neck region to behind the ear to exit at the burr hole site, and connected to the catheter.
- The peritoneal catheter is undermined subcutaneously over the paravertebral muscles and exits over the lateral abdomen.
- The peritoneum is incised, and the catheter is slowly coiled into the abdomen and sutured in place.
- The entire system is connected and checked for appropriate drainage function.

**Removal of the Inciting Obstructive Cause**
Procedure depends on the cause and may range from chemotherapy to surgical intervention to radiation therapy.

**Supportive Treatment**
- Standard postoperative care with appropriate pain relief (e.g., hydromorphone, 0.1 mg/kg IV q4h or PRN).
- Prophylactic broad-spectrum, bactericidal antibiotic therapy is recommended at the initiation of surgery and for a minimum of 5 days after any shunting procedure.
- Nutritional support is important. Feeding is recommended within 24 hours of the procedure.
- Behavioral training/modification—Some dogs with congenital hydrocephalus stabilize over time and may become quality pets with appropriate environmental adaptations by the owner and pet.

**Patient Monitoring**
- Serial neurologic examinations to evaluate patient progress and/or benefit of therapy.
- **Electroencephalography** may be a noninvasive, sensitive, and reliable method to evaluate improvement of brain function.
- **Cranial imaging** is important if shunting has been performed to determine location of shunt placement.

**V-P shunt care:**
- **Immediate postoperative period**—Heart rate and blood pressure monitoring. Bradycardia and systemic hypertension are indications of elevated ICP.
- **Monitor body temperature**—Fever is a common initial sign of shunt infection.
- If infection is suspected, CSF can be collected from the reservoir component of the valve system, analyzed, and cultured.

**Home Management**
- Seizure management as needed.
• Shunt care
  - Exercise should be restricted for the first 30 days, followed by careful observation to avoid excessive fighting/playing with other dogs.
  - Neck leads and chains are to be avoided. A chest harness should be used at all times.
  - Owner should be aware of early signs of infection and be instructed to monitor rectal temperature as needed to determine presence of fever.

Milestones/Recovery Timeframes
• First 24 hours—Improved alertness.
• First 7 days—Improved behavior.
• First 30 days—Improved behavior without recurrence of signs.
• Beyond 30 days—Stabilization of signs.

TREATMENT OPTIONS

Medical Management to Reduce ICP
• Prednisone—0.25–0.5 mg/kg PO bid for 7–14 days initially, then taper to an acceptable daily to every other day dose (around 0.1 mg/kg/day) as needed.
• Prednisolone sodium succinate—10 mg/kg IV q6h for 24 hr.
• Mannitol (20%)—1 g/kg IV over 20 min without concurrent fluid administration for treatment of HPH. An additional two doses (0.25–0.5 g/kg) may be given within 24 hr.
• Furosemide—0.7–1.0 mg/kg IV once at termination of the mannitol therapy for treatment of HPH. May be repeated twice in 24 hr.
• Acetazolamide—10 mg/kg PO q8–12h for chronic medical management of NPH (dogs only).
• Hyperventilation to a Paco2 of 25–35 mmHg in conjunction with barbiturate general anesthesia.

Surgical Management to Reduce ICP
• Acute, temporary procedure:
  - Elsberg ventricular catheter, 4 in., 5–7 Fr (Codman®, Johnson & Johnson)
• Chronic, permanent procedure:
  - Codman® Hakim Precision Valve, medium- to low-pressure range (70 ± 10 mmH2O) ventriculoperitoneal shunt system (Codman®, Johnson & Johnson)

Antiepileptic Drug Therapy
• Diazepam—0.5 mg/kg IV or 1–2 mg/kg per rectum to stop seizure activity.
• Phenobarbital—2.5 mg/kg PO bid for duration of disease as needed to prevent seizure onset.
• Potassium bromide—40 mg/kg PO daily with food; can be divided bid.

STANDARDS OF CARE: EMERGENCY AND CRITICAL CARE MEDICINE

Favorable Criteria
• Improved alertness and behavior.
• Improved neurologic examination.
• Reduced or no seizure activity.

Unfavorable Criteria
• Failure to exhibit improved behavior and alertness after medical and/or shunt therapy.
• Increasing neurologic deficits.
• Increasing seizure frequency.
• Shunt complications:
  - Undershunting: Due to disconnection, kinking, or blockage. Causes for the latter include obstruction by choroid plexus or cellular debris along the entire catheter.
  - Overshunting: Possible slit ventricle syndrome with associated brain collapse and intracranial and/or intraventricular hemorrhage.

PROGNOSIS

The prognosis for an animal’s return to a quality life depends on a number of variables, including the type and severity of the hydrocephalus, amount of brain atrophy, underlying etiology, and time course of interventional therapy as related to disease onset. As with many other neurologic disorders, the earlier the onset of treatment, the better the chance for a positive clinical outcome. In general, many dogs with congenital NPH hydrocephalus can lead a high-quality life (i.e., can interact well with owners and acclimate to their home environment). Residual signs, such as seizures, may persist. The hydrocephalus can also progress slowly over time despite medical treatment. Animals with acquired obstructive HPH will often exhibit a dramatic improvement after a V-P shunting procedure, with many dogs returning to almost normal function. However, relapse of clinical signs may occur if the underlying etiology progresses or if complications from shunt placement occur.
RECOMMENDED READING


