Congenital Hydrocephalus

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KEYWORDS

• Hydrocephalus • Brain • Cerebrospinal fluid • Ventricular system
• Ventriculoperitoneal shunt

KEY POINTS

- There are several types of hydrocephalus, which are characterized based on the location of the cerebrospinal fluid (CSF) accumulation.
- Physical features of animals with congenital hydrocephalus may include a dome-shaped skull, persistent fontanelle, and bilateral ventrolateral strabismus.
- Medical therapy involves decreasing the production of CSF.
- The most common surgical treatment is placement of a ventriculoperitoneal shunt.
- Postoperative complications may include infection, blockage, drainage abnormalities, and mechanical failure.

INTRODUCTION

A current definition of hydrocephalus is an active distension of the ventricular system of the brain that results from inadequate movement of cerebrospinal fluid (CSF) from the point of production within the ventricles to its point of absorption.[1] Congenital hydrocephalus typically occurs because of an interruption of CSF flow or defective CSF absorption; hydrocephalus is rarely caused by an increase in CSF production. CSF is formed primarily by the choroid plexus in the lateral, third, and fourth ventricles at a rate of 0.047 mL/min in dogs and 0.017 mL/min in cats.[2] Additional CSF is secreted by the ependymal lining, the external pial-glial membrane on the surface of the brain, and by the blood vessels in the pia-arachnoid.[3,4] Production of CSF is independent of CSF hydrostatic pressure and occurs at a constant rate; however, it depends on osmotic pressure. Normal CSF flow begins in the lateral ventricles and travels through the interventricular foramen to the third ventricle, and from this point it enters the mesencephalic aqueduct to emerge in the fourth ventricle (**Fig. 1**). From the fourth ventricle, CSF exits via the lateral apertures to enter the subarachnoid space. Movement of CSF through the ventricular system is caused by pumping of blood in the choroid plexus. The bulk of CSF absorption takes place at the arachnoid villi and to a lesser degree via venous and lymphatic drainage around spinal and

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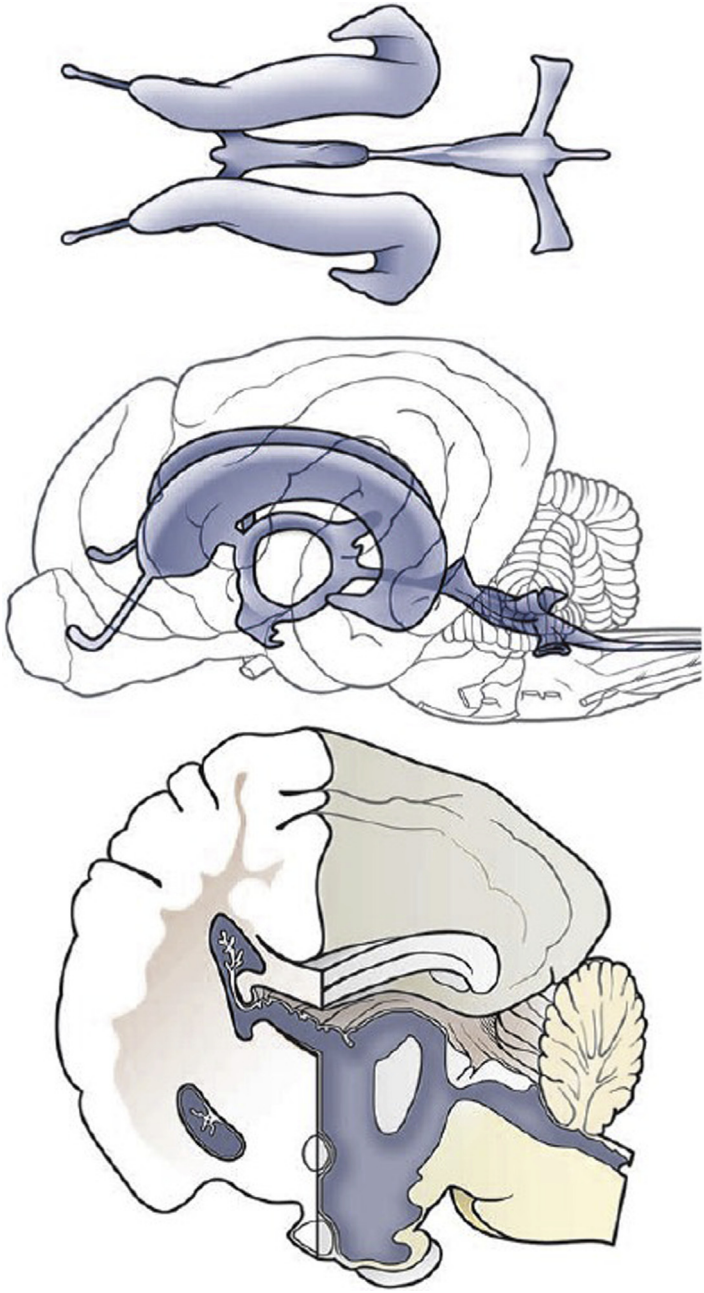

Fig. 1. The normal ventricular anatomy of the canine brain. (*From* Dewey CW, Marino DJ. Congenital brain malformations. In: Tobias KM, Johnston SA, editors. Veterinary surgery, small animal. Philadelphia: Elsevier; 2012. p. 518; with permission.)

cranial nerves. The arachnoid villi are projections of subarachnoid space into the lumen of the venous sinus. The section of the villus that is within the sinus acts as a valve that allows CSF to flow into the lumen of the venous sinus when CSF pressure is higher than venous pressure but collapses when venous pressure is higher, preventing blood from entering the subarachnoid space.[4,5]

TYPES OF HYDROCEPHALUS

Hydrocephalus can be divided into congenital and acquired. There are various types of hydrocephalus, which are characterized based on the location of the CSF accumulation.

- Internal hydrocephalus is the accumulation of CSF within the ventricular system.
- External hydrocephalus is the accumulation of CSF within the subarachnoid space.
- Communicating hydrocephalus is when the CSF in the ventricular system communicates with the subarachnoid space, resulting typically from an obstruction beyond the fourth ventricle.
- An obstructive (or noncommunicating) hydrocephalus is ventricular dilation resulting from a lesion causing obstruction of CSF flow before entering the subarachnoid space.
- A compensatory hydrocephalus can result from loss of central nervous system parenchyma, whereby there is an increase in CSF volume that occupies the space formerly taken up by the lost parenchyma.

PATIENT EVALUATION OVERVIEW

Dogs and cats with congenital hydrocephalus may have signs from birth; however, more commonly signs become apparent in the first few months of life. The rate of clinical progression of congenital hydrocephalus is variable, and some animals may not develop clinical signs of encephalopathy until adulthood. Congenital hydrocephalus is overrepresented in toy breed dogs. Breeds found to be at a higher risk include the Maltese, Yorkshire terrier, English bulldog, Chihuahua, Lhasa apso, Pomeranian, toy poodle, cairn terrier, Boston terrier, pug dog, and Pekingese.[6] The most commonly reported malformation is a stenotic mesencephalic aqueduct, which is often seen with a malformation of the mesencephalon affecting the rostral colliculi and rarely the caudal colliculi.[5] Another potential mechanism is the hydrodynamic theory, which proposes that hydrocephalus develops on account of decreased intracranial compliance, resulting in increased capillary pulse pressure. A pulsatile pressure gradient develops that is directed from the cerebral tissue toward the lateral ventricles. The rebound pressure from the pulsatile gradient, along with the hyperdynamic CSF flow in the mesencephalic aqueduct, results in enlargement of the ventricular system. This enlargement results in congenital hydrocephalus that can develop in the face of normal intracranial pressure.[7] Often a specific cause for the hydrocephalus is not obvious at the time of initial evaluation.

PHYSICAL EXAMINATION FINDINGS

Common physical characteristics of hydrocephalic patients include a large, dome-shaped head, fontanelles or larger calvarial defects, and bilateral ventrolateral strabismus (sun-setting of the eyes) (**Fig. 2**). The strabismus may be the result of either orbital skull malformations or vestibular dysfunction. Dogs and cats with congenital hydrocephalus often show evidence of developmental delay and are smaller than

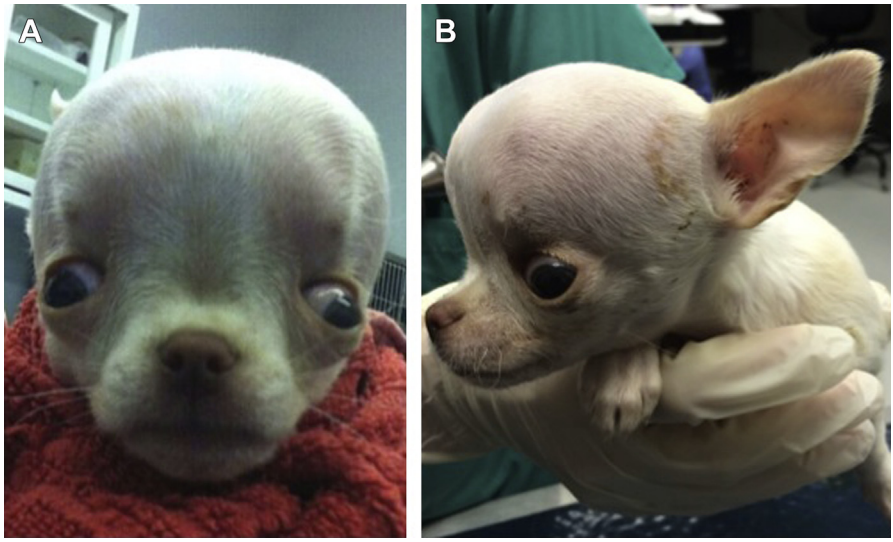

Fig. 2. A frontal (*A*) and lateral (*B*) view of a Chihuahua with congenital hydrocephalus demonstrating an enlarged dome-shaped skull and ventrolateral strabismus.

normal. Clinical signs of neurologic dysfunction usually reflect a forebrain disorder and may include obtundation, behavioral abnormalities, difficulty with training, decreased vision or blindness, circling, pacing, and seizure activity. Concurrent congenital abnormalities of the brain (eg, intracranial arachnoid cyst, Dandy-Walker syndrome, Chiari-like malformation) may also occur in hydrocephalic dogs. Progressive neurologic dysfunction over weeks to months is often a reason for pursuit of medical and/or surgical intervention.

DIAGNOSIS

Diagnosis of congenital hydrocephalus is based on a combination of characteristic clinical features, imaging of the brain to demonstrate ventriculomegaly, and the absence of other causes of encephalopathy. It is important to note that the terms *ventriculomegaly* and *hydrocephalus* are not synonymous. Although enlarged ventricles are a feature of hydrocephalus, not all animals with ventriculomegaly have hydrocephalus. Asymmetric and symmetric enlargement of the lateral ventricles can be seen in neurologically normal animals. In one study looking at normal beagle-type dogs, enlargement of one or both ventricles was seen in 46.7% of the dogs.[8] In another study, the prevalence of asymmetry of the lateral ventricles in a group of 100 neurologically normal dogs was 38%.[9] It is important to use clinical signs and to identify other features on imaging that can help support this diagnosis, such as periventricular hyperintensities on MRI, a site of obstruction, and ventricular enlargement that cannot be entirely attributed to cortical atrophy, to name a few.

IMAGING MODALITIES

Ultrasound

If a dog presents with a persistent fontanelle, then ultrasound may be a safe, minimally invasive modality to image the ventricles. The lateral ventricles would appear enlarged and filled with anechoic fluid and lined by a thin wall. The primary benefit is this can often

be done with no sedation. More advanced imaging techniques should be performed following ultrasound evaluation if medical or surgical treatments are to be pursued.

Computed Tomography Scan

Computed tomography (CT) allows visualization of the entire ventricular system. It may be possible to identify a site of obstruction or the morphology of the ventricular system may suggest the location of an obstruction. This modality would also allow visualization of acute hemorrhage. CT can also be used postoperatively to confirm shunt placement (**Fig. 3**).

MRI Scan

MRI will allow for the best resolution and detailed evaluation of the brain parenchyma (**Fig. 4**). Periventricular hyperintensities in the white matter can be seen and often indicate interstitial edema (**Fig. 5**). MRI will also be more useful for identifying concurrent abnormalities (eg, Chiari-like malformation).

MEDICAL TREATMENT

Medical management of hydrocephalus is directed at decreasing the production of CSF. This treatment may be performed in cases when surgery is not an option or in an attempt to stabilize ventricular size and clinical signs until a ventriculoperitoneal shunt can be placed.[10] Antiepileptic drugs may also need to be given if patients are experiencing seizures. Medical therapy may stabilize or improve signs in the short-term, but often it is not successful in the long-term.

Diuretics are used to reduce the production of CSF. Acetazolamide, a carbonic anhydrase inhibitor, has been shown to decrease CSF production by the choroid plexus in both the dog and cat.[11,12] Although acetazolamide can reduce CSF production, this does not always correspond with a reduction in intracranial pressure. In humans with idiopathic normal-pressure hydrocephalus, low-dose acetazolamide has been associated with a reduction in periventricular white matter hyperintensities

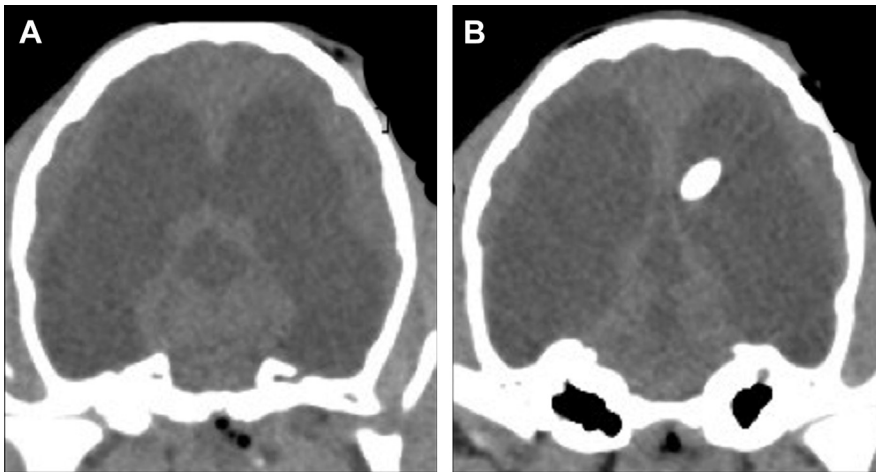

Fig. 3. Preoperative CT scan showing enlarged lateral ventricles in a young dog with congenital hydrocephalus (*A*) and postoperative image showing placement of the ventriculoperitoneal shunt in the lateral ventricle (*B*). (*Courtesy of* Dr Emma Davies, Cornell University Hospital for Animals, Ithaca, NY.)

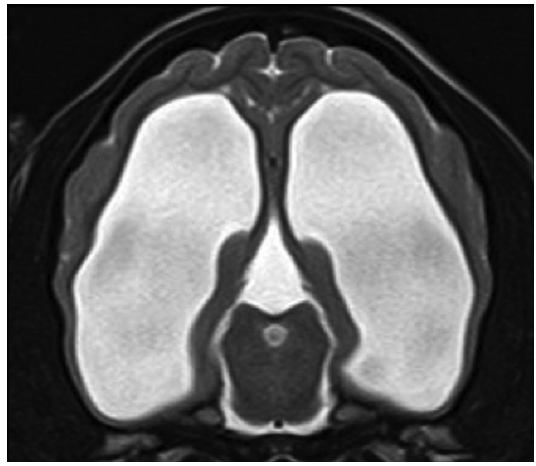

Fig. 4. Transverse T2-weighted image showing dilated lateral ventricles from a hydrocephalic puppy. (*Courtesy of* Dr Emma Davies, Cornell University Hospital for Animals, Ithaca, NY.)

seen on MRI.[13] Furosemide is a loop diuretic that acts mainly by inhibiting sodium reabsorption in the nephron at the thick ascending limb of the loop of Henle.[14] Furosemide has also been shown to decrease CSF production and intracranial pressure in rabbits.[15] Omeprazole is a proton pump inhibitor that has been shown to decrease CSF production and in one study was found to decrease production by 26%.[16,17]

Glucocorticoids are commonly used in patients with hydrocephalus, although there is little published information available regarding their efficacy in these patients. Typically an antiinflammatory dose is used initially; once clinical signs have improved, the medication is tapered to the lowest dose that still controls clinical signs.

Hyperosmolar therapy with agents such as mannitol is occasionally used when there is evidence of intracranial hypertension.[10] Mannitol can reduce intracranial pressure by decreasing blood viscosity, which promotes reflex vasoconstriction; mannitol also produces an osmotic gradient that draws fluid from the brain parenchyma into the

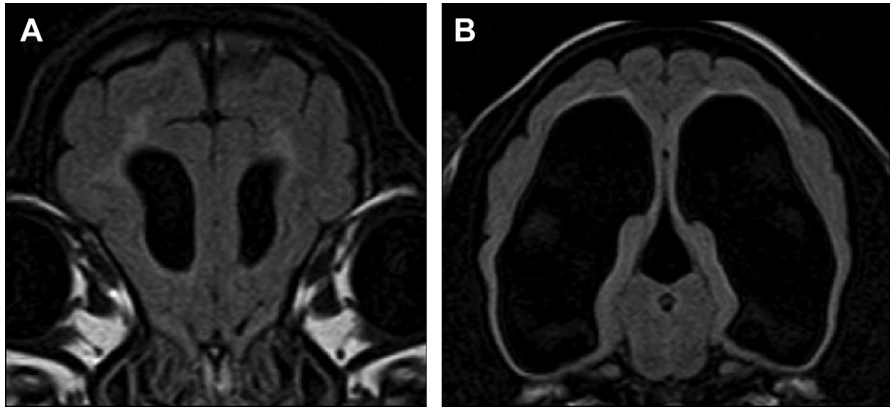

Fig. 5. Transverse fluid attenuated inversion recovery images at the level of the rostral cerebrum (*A*) and mesencephalic aqueduct (*B*) showing dilated lateral ventricles and periventricular hyperintensities. (*Courtesy of* Dr Emma Davies, Cornell University Hospital for Animals, Ithaca, NY.)

vascular space.[18,19] Side effects of mannitol administration may include hypovolemia, electrolyte imbalances, and acute renal failure, so follow-up therapy with isotonic crystalloids may be warranted. Mannitol may result in an initial clinical response but is not suited for long-term therapy, and the response is likely to be transient.[20]

The potential side effects of long-term corticosteroid and/or diuretic therapy need to be considered when deciding about the best course of action for treating congenital hydrocephalus. Electrolyte depletion (particularly potassium) and dehydration are concerns when using diuretics for prolonged time periods, particularly when used in combination with corticosteroids.

Ultrasound-guided ventriculocentesis via the lateral ventricle can be performed in some cases with a persistent fontanelle. This technique can be used to provide temporary relief in clinical patients and also to determine a potential response to placement of a ventriculoperitoneal shunt (**Table 1**).

INDICATIONS FOR SURGERY

There is no clear outline that describes when an animal should have a ventriculoperitoneal shunt placed. Surgical treatment is generally recommended when an animal is showing worsening clinical signs or shows no evidence of improvement or deteriorates when being treated medically. It is important that surgery not be performed in patients with preexisting skin infections at the sites of surgery, systemic infection, or an abdominal infection.[4] Surgical candidates should not have other brain malformations that could be contributing to the neurologic signs that would not get addressed by shunt placement. It is also important to recognize cases that would not benefit from placement of a ventriculoperitoneal shunt, such as those with chronic irreversible changes and those with ventricular enlargement secondary to other causes (eg, cerebral necrosis).[4]

SURGICAL TREATMENT

The use of ventriculoperitoneal shunts is the mainstay for surgical treatment of congenital hydrocephalus. The 4 primary shunt components are a ventricular catheter, a reservoir, a valve, and a distal (peritoneal) catheter. To place a shunt, a burr hole is made in the calvarium lateral to the nuchal crest. The dura mater is incised, and a path for the shunt is made using a needle or stylet gently inserted into the cerebral cortex at the level of the lateral ventricle. The ventricular catheter is placed in the lateral ventricle and is anchored to the skull (**Fig. 6**). The distal portion of the catheter is tunneled subcutaneously and then inserted into the peritoneum via a paralumbar approach. A sufficient amount of additional tubing should be placed in the abdomen to allow for growth of the patients. A catheter can also be placed in the atrium; however, this is technically more challenging, and the jugular vein can be too small for catheter placement in small dogs. It is important to do postoperative imaging to confirm appropriate placement of the shunt (**Figs. 7** and **8**).

Table 1
A list of medications that have been shown to aid in decreasing CSF production

Drug	Class	Dosage
Prednisone	Glucocorticoid	0.25–0.5 mg/kg PO q12h, followed by a taper
Omeprazole	Proton pump inhibitor	1 mg/kg PO q24h
Acetazolamide	Carbonic anhydrase inhibitor	10 mg/kg PO q8h
Furosemide	Loop diuretic	1 mg/kg PO q24h

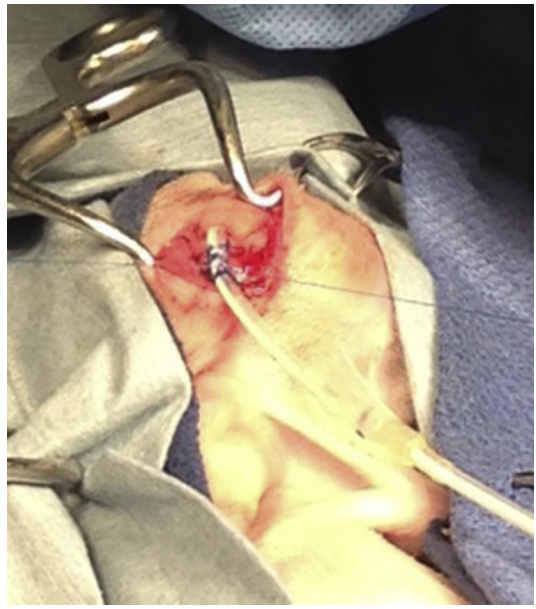

Fig. 6. Intraoperative image of shunt placement in the lateral ventricle for treatment of congenital hydrocephalus. The shunt has been anchored to the calvarium with a Chinese finger trap pattern.

Most of the valves in use are differential pressure valves that will open when the difference in pressure across the valve is greater than the preset opening pressure, which will allow outflow of CSF. When the pressure difference decreases below the set point, the valve closes, stopping movement of CSF.[21,22] Externally programmable valves are also available, which allow the clinician to noninvasively adjust the opening pressure by use of a magnetic device. Depending on the variety, some of the programmable valves can be reset in the MRI machine, whereas others have a locking mechanism in place to prevent this. Most valves are compatible with MRI machines up to 3 T; however, evaluation after imaging and readjustment of the settings may be needed to ensure that correct opening pressure is maintained.[23,24]

SURGICAL COMPLICATIONS

Ventriculoperitoneal shunting is a well-established practice in the treatment of congenital hydrocephalus. Most patients tolerate the surgery and show improvements postoperatively. However, there are several well-known complications that can be seen with this procedure.

Infection

Shunt infections are seen in approximately 8% of cases in people and are typically identified within 2 months of surgery.[21] Treatment of an infection often requires removal of the shunt, culturing the organism, and instituting treatment based on sensitivity results. MRI features of a shunt-associated infection have been outlined in a dog. These features included T2-weighted and fluid-attenuated inversion recovery hyperintensity of the ventricular lining and marked contrast enhancement of the ependymal layer on T1-weighted postcontrast images.[25]

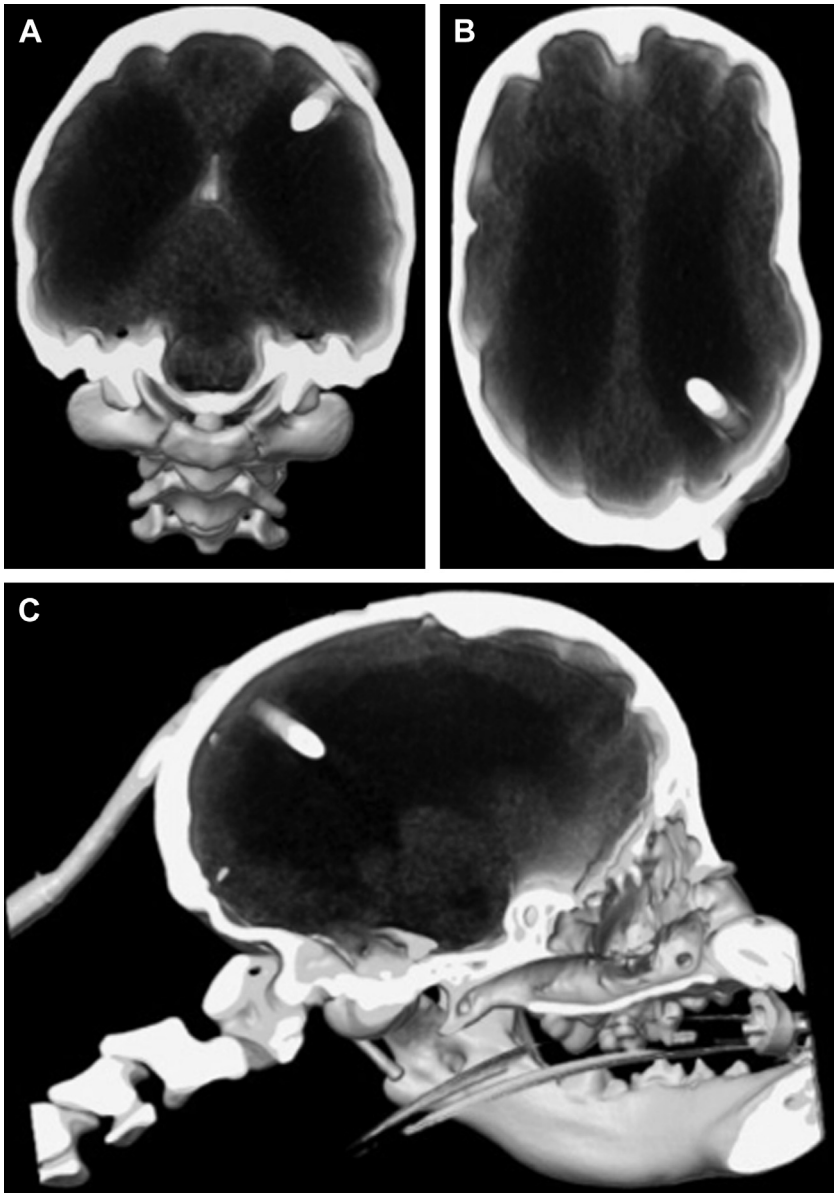

Fig. 7. Postoperative 3-dimensional CT reconstruction images in transverse (*A*), dorsal (*B*), and sagittal (*C*) planes showing shunt placement in the lateral ventricle. (*Courtesy of* Dr Emma Davies, Cornell University Hospital for Animals, Ithaca, NY.)

Blockage

Various components of the shunt can become blocked. Blockage can occur as a result of debris (tissue, hemorrhage, highly cellular CSF) occluding the catheter, obstruction by choroid plexus, the tip of the catheter becoming lodged within or against the brain parenchyma, and the peritoneal portion of the catheter can become occluded or similarly embedded within or against tissue. It is also possible for the

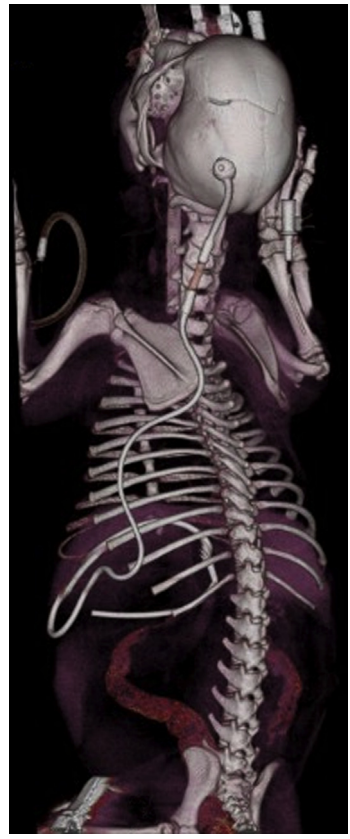

Fig. 8. Postoperative 3-dimensional CT dorsal plane reconstruction image showing placement of the ventriculoperitoneal shunt. (*Courtesy of* Dr Emma Davies, Cornell University Hospital for Animals, Ithaca, NY.)

shunt to become kinked, which would prevent adequate drainage. Blockage may require surgical exploration and replacement of the faulty component.

Drainage

It is possible that overdrainage can cause the ventricles to collapse. This collapse can result in CSF that accumulates between the brain and skull and can also cause subdural hematoma formation secondary to tearing of the vasculature. In these cases, a valve with a higher opening pressure or a device with a programmable valve may be needed. Underdrainage can result when the catheter system is blocked, becomes disconnected, or is kinked.

Mechanical Failure

Shunt malfunction can be the result of breakage, migration, or disconnection of one or multiple components of the shunt.

PROGNOSIS

Overall the success rate for dogs treated with shunting falls in the range of 72% to 100%.[26–29] In one recent veterinary study evaluating 36 cases of congenital

hydrocephalus in dogs and cats, it was found that there was an overall improvement in clinical signs in 72% of the animals. Twenty-two percent of the animals developed postoperative complications, and 36% of the animals died of hydrocephalus-related complications or were euthanized.[27] As in people, this study found that most complications were seen within a few months following surgical intervention. In another study of 14 dogs, ventriculoperitoneal shunting was successful in improving neurologic signs in most dogs, and postoperative complications were seen in 29% of the patients; but these complications could be addressed either medically or surgically.[28] In a study of 12 dogs with hydrocephalus that underwent ventriculoperitoneal shunting, all showed signs of neurologic improvement after shunt placement; however, 25% of the dogs were euthanized because of a lack of sustained improvement or skull pain. The other animals had sustained improvement, and one did require revision surgery.[29] In a human study looking at ventriculoperitoneal shunt placement outcomes in infants, it was found that the risk of shunt failure was higher in infants less than 1 month of age at the time of shunt placement.[30] In another human study of 198 pediatric patients with hydrocephalus, it was found that 8.6% of patients experienced shunt infections and they were more likely to be underweight compared with those without infection. In this study the mean interval between shunt placement and infection was 1.83 $\pm$ 1.25 months and coagulase-negative *Staphylococcus* was the most commonly encountered pathogen.[31] In a human study of 333 consecutive ventriculoperitoneal shunts placed in pediatric patients, it was found that 35 shunts (10.5%) were infected and that infection occurred at a median of 1 months after shunt placement.[32] As in the previously mentioned study, the most common causative agent was coagulase-negative *Staphylococcus*. This study also found that an independent risk factor for infection was having surgery before 1 year of age.[32] Similar investigations looking at risk factors have not been performed in veterinary medicine, but it is possible that age could be a risk factor in veterinary patients as well, for similar reasons, such as less developed immune systems, immaturity of the skin barrier, and the population of bacterial flora.[30,32]

It is important to have a detailed discussion with the owners regarding the potential complications associated with this surgery and the possible need for a revision surgery should these issues be encountered postoperatively. As in human medicine, complications are most commonly seen in the first few months following surgery, so frequent reevaluation is important in the early postoperative period.

Presently, placement of a ventriculoperitoneal shunt in patients with congenital hydrocephalus is still the best treatment option in those animals that are deemed to be appropriate surgical candidates.

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