Hydrocephalus in canine and feline patients

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Introduction:
Hydrocephalus is the excessive accumulation of cerebrospinal fluid (CSF) within the ventricular system of the brain and can be caused by several mechanisms including an obstruction of normal CSF flow, decreased CSF absorption, or excessive production of CSF. There are several classification systems used to describe the underlying cause of hydrocephalus. However, in canine and feline patients it is most useful to think of hydrocephalus in terms of two main categories: 1) congenital hydrocephalus and 2) obstructive hydrocephalus. Rarely, a third type of hydrocephalus caused by excessive CSF production can be seen in patients with a choroid plexus tumor.

Congenital Hydrocephalus:
Congenital hydrocephalus is most often first recognized at an early age and most commonly occurs in small and toy breed dogs. When imaging (MRI) is performed the hydrocephalus is often most apparent in the cerebral hemispheres and is characterized by enlarged lateral ventricles and decreased cerebral cortical thickness. Most often the underlying cause of the congenital hydrocephalus is not apparent even after extensive diagnostics are performed.

Obstructive Hydrocephalus:
CSF flows primarily in a cranial to caudal direction, traveling out of the ventricular system, subarachnoid space, and collection of cisterns in the cranial cavity and into the spinal cord via the subarachnoid space and central canal of the spinal cord. A blockage in the flow of CSF is a concern because production of CSF is largely independent of intraventricular pressure. Thus, the absorption of CSF cannot compensate for an obstruction of normal flow and continued production, and the resulting increased pressure results in pathologic changes to the brain parenchyma in addition to marked clinical manifestations. Common lesions that can result in obstructive hydrocephalus include neoplasia, infectious etiologies, and immune-mediated etiologies.

Diagnosis:
The diagnosis of congenital hydrocephalus is most commonly made based on a patient's clinical signs, signalment, and ultimately on MRI findings. Ultrasound and electroencephalography (EEG), although used less commonly, can also aid in the diagnosis.

Clinical signs of congenital hydrocephalus are variable, although most often signs reflect forebrain dysfunction. Signs can include a depressed to obtunded mentation, pacing and circling, seizures, and behavioral abnormalities, among others. Sometimes patients have a dome-shaped head, open fontanelle, or ventrolateral

strabismus. Classic breeds predisposed to congenital hydrocephalus include small and toy breed dogs such as the chihuahua and Yorkshire terrier.

Clinical signs seen with obstructive hydrocephalus are more variable and are dependent on the level of the obstruction. For example, an obstruction at the level of the caudal brainstem may cause primarily vestibular signs if there is excessive buildup of CSF in the fourth ventricle.

MRI is an extemely useful tool in the diagnosis of both congenital and obstructive hydrocephalus. However, in cases of congenital hydrocephalus there is no single objective MRI parameter that can be measured to make a diagnosis. Complicating matters is the fact that many small and toy breeds have large ventricles (ventriculomegaly) that are not necessarily the cause of clinical signs. In such cases a diagnosis is based not only on MRI findings but also on patient signalment, history, and by ruling out other conditions such as metabolic (e.g. hepatic encephalopathy) and degenerative (e.g. lysosomal storage diseases) etiologies that can produce overlapping clinical signs.

Making the diagnosis of obstructive hydrocephalus based on MRI is more straightforward. MRI findings consistent with obstructive hydrocephalus include enlarged rounded ventricles and the presence of interstitial edema, a transependymal movement of CSF best seen on FLAIR images. In addition, the presence of brain herniation, which infers increased intracranial pressure, lends further support to the diagnosis of obstructive hydrocephalus. In many cases MRI also allows for the determination of the underlying cause of the obstruction.

Treatment:
There is significant overlap between treaments for congenital and obstructive hydrocephalus. Treatment of both types of hydrocephalus can be divided into medical and surgical therapies. Medical treatments are aimed at reducing production of CSF and can include administration of prednisone, diuretics (acetazolamide, furosemide), and omeprazole. Additionally, mannitol, an osmotic diuretic, is effective at reducing CSF production and is useful in the acute setting to reduce intracranial pressure.

In human medicine, and increasingly in veterinary medicine, hydrocephalus is often viewed as a surgical disease. Treatment centers around diverting CSF out of the ventricular system to an alternate location for absorption. In feline and canine patients this diversion is most often accomplished by placing a ventriculoperitoneal (VP) shunt, a device that re-routes CSF from the central nervous system into the peritoneal cavity. Most often the VP shunt is inserted into a lateral ventricle and tunneled subcutaneously to the abdomen where it is inserted into the peritoneal cavity. The most common complications of VP shunts include infection, obstruction, and intracranial hemorrhage.

In cases of obstructive hydrocephalus therapy also involves treatment of the underlying cause. This may include surgery, radiation therapy, and/or chemotherapy in cases of neoplasia; antibiotics or antifungals in the cases of infectious etiologies; or immunosuppressants in the case of immune-mediated etiologies.

Prognosis:
For obstructive hydrocephalus, the prognosis is extremely dependent on the cause of the obstruction and the therapies pursued. For example, a patient with obstructive hydrocephalus secondary to an infectious process often has a good prognosis if the underlying infection is treated successfully and the CSF buildup relieved either medically or surgically. Often these patients require short-term therapies (e.g. VP shunt) to treat life-threatening increased intracranial pressure while longer term therapies (e.g. antibiotics or antifungals) have time to treat the underlying cause. In contrast, the prognosis for a patient with hydrocephalus secondary to a primary aggressive neoplasm may be more guarded even with aggressive treatment.

In cases of congenital hydrocephalus the prognosis is dependent on the amount of remaining functional brain tissue (e.g. remaining cerebral cortex). If clinical signs are severe (e.g. the patient is obtunded or stuporous) then the prognosis is often guarded. However, if clincial signs are mild and slowly progressive then often times patients may benefit from treatment.

Conclusion:
It is important to accurately diagnose the type of hydrocephalus (congenital vs. obstructive, while remembering that there are less common causes of hydrocephalus), which is best done by synthesizing patient signalment, history, and diagnostic test results. Once a diagnosis is made it is essential to commence therapies (medical and surgical options) directed at treating the hydrocephalus directly and, in the case of obstructive hydrocephalus, to consider treatments (surgery, radiation therapy, chemotherapy, antibiotics, antifungals, immunosuppressants, etc.) directed at the underlying condition (neoplasia, infectious, immune mediated, etc.). Lastly, patients selected for surgical intervention (i.e. placement of a ventriculoperitoneal shunt) must be chosen carefully as prognosis can vary significantly from case to case.

Selected References:

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