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Intracranial Arachnoid Cysts in Dogs

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Abstract: Intracranial arachnoid cyst (IAC) is an infrequently reported developmental disorder seen primarily in small-breed dogs. It usually occurs in the caudal fossa, in the region of the quadrigeminal cistern. Although still considered uncommon, IAC is being recognized more frequently in veterinary medicine, coinciding with the increased availability of magnetic resonance imaging. In this article, clinical information from previously reported cases of canine IAC is combined with additional case information from our hospitals. Similar to IAC in people, it is thought that canine IAC is often an incidental finding. When IAC is responsible for neurologic disease in dogs, generalized seizures and cerebellovestibular dysfunction are the most common clinical presentations. Medical therapy of IAC focuses on management of increased intracranial pressure and seizures, if the latter are part of the clinical complaints. Surgical therapy of IAC involves either cyst fenestration or shunting the excess fluid to the peritoneal cavity.

Intracranial arachnoid cyst (IAC), also called *intracranial intraarachnoid cyst* and *quadrigeminal cyst*, is a developmental brain disorder in which cerebrospinal fluid (CSF) is thought to accumulate within a split of the arachnoid membrane.¹ Although IACs have been reported to occur in several locations in humans, all reported canine cases have been in the

caudal fossa.²⁻¹¹ Because IAC is typically associated with the quadrigeminal cistern in dogs, these accumulations of fluid are often called *quadrigeminal cysts* in this species and have a characteristic appearance on magnetic resonance imaging (MRI) scans^{4-6,10,11} (FIGURE 1). IACs account for 1% of all intracranial masses in people and are often considered incidental findings.¹²⁻¹⁴

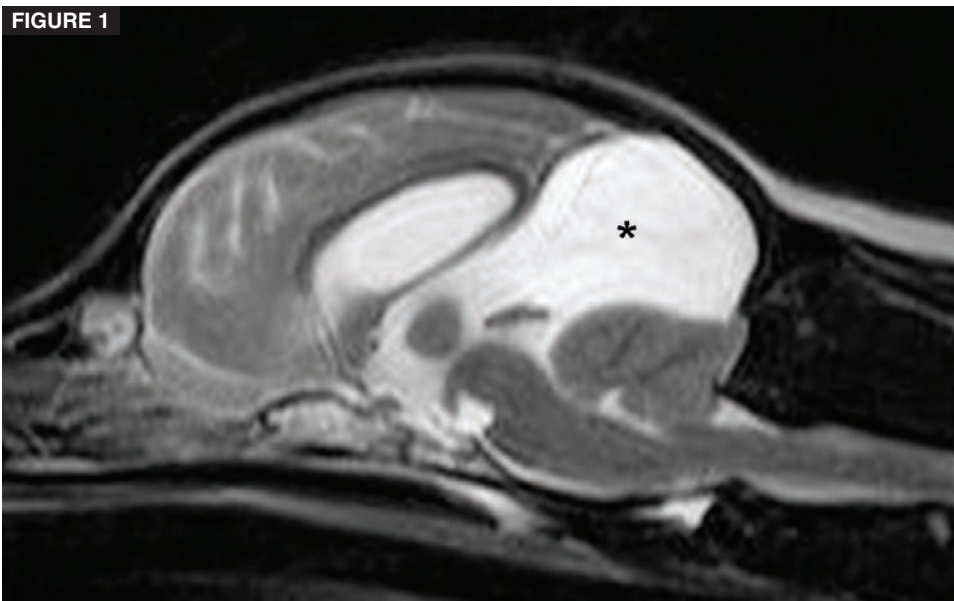


FIGURE 1 Midsagittal T2-weighted image of a dog's brain demonstrating the characteristic appearance of an IAC in this species. The asterisk is in the center of the cyst. (Reprinted with permission from Dewey CW. Encephalopathies: disorders of the brain. In: Dewey CW, ed. *A Practical Guide to Canine and Feline Neurology*. 2nd ed. Ames, Iowa: Wiley-Blackwell; 2008:115-120.)

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There are 10 clinical reports of IAC in dogs in the veterinary literature.²⁻¹¹ This review combines these reported cases with three additional cases from our hospitals to present information regarding 56 dogs with IAC. Most reported cases of IAC in dogs are in small breeds, with a predominance of brachycephalic animals.²⁻¹¹ Shih tzus may be overrepresented.^{11,15} Male sex also appears to be a predisposing factor. Clinical signs attributable to IAC in dogs are most often related to cerebral or cerebellar compression by the cyst; generalized seizures and central vestibular dysfunction are most commonly noted.¹⁵ Similar to human IACs, a large proportion of reported IACs in dogs were suspected to be incidental.^{4-6,11} Medical and surgical options are available to treat IAC in dogs.

tissue called the *perimedullary mesh*; this tissue eventually becomes the pia and arachnoid layers of the meninges. In normal development, pulsatile CSF flow from the choroid plexuses is thought to divide the perimedullary mesh into the pia and arachnoid layers, effectively creating the subarachnoid space. It is postulated that some aberration of CSF flow from the choroid plexuses during this stage of development forces a separation within the forming arachnoid layer, eventually leading to the creation of an IAC.^{1,12} The intrarachnoid location of IACs has been demonstrated via light and electron microscopy in people.¹ Depending on whether these cysts communicate with the subarachnoid space or the ventricular system, they are sometimes referred to as *communicating* or *noncommunicating*.¹

The mechanisms by which an IAC continues to expand with fluid are unknown, but several theories have been proposed.^{1,12} There is evidence that arachnoid cells lining the IAC may have secretory capacity.^{1,12,16} Fluid may also move into the cyst via an osmotic pressure gradient. However, considering that the fluid within the IAC is nearly identical to CSF, this theory is unlikely.^{1,12} In addition, there have been documented cases in people in which small slits exist between the IAC and the subarachnoid space; these slits act as one-way valves, diverting CSF into the cyst during systole and preventing its return to the subarachnoid space during diastole.^{1,12,17}

Clinical Features

Most reported dogs with IAC have been small breeds, and many had brachycephalic conformation. The following information was obtained by combining the IAC cases reported in the literature with three additional cases from our hospitals. The breed distribution of these 56 dogs is listed in **BOX 1**. Approximately 66% of the dogs (37 of 56) were male.²⁻¹¹

There is a wide age range at clinical presentation for dogs with IAC (2 months to 12 years), with an approximate average age of 4 years. The most common clinical signs (**BOX 2**) seen with IAC are reflective of forebrain or central vestibular (cerebellovestibular) dysfunction. Other reported clinical signs include neck pain, paraparesis, and tetraparesis.²⁻¹¹

Pathogenesis

IACs are believed to represent a developmental abnormality caused by an aberrant split in the arachnoid membrane during embryogenesis.^{1,12} The developing neural tube is surrounded by a loose layer of mesenchymal

BOX 1

Breed Distribution of Reported Intracranial Arachnoid Cysts in Dogs

| | |
|---|---------|
| ▶ Shih tzu | 15 dogs |
| ▶ Maltese | 4 |
| ▶ Pug | 4 |
| ▶ Lhasa apso | 4 |
| ▶ Cavalier King Charles spaniel | 4 |
| ▶ Yorkshire terrier | 4 |
| ▶ Chihuahua | 3 |
| ▶ Staffordshire bull terrier | 3 |
| ▶ Bulldog | 3 |
| ▶ Pekingese | 2 |
| ▶ West Highland white terrier | 2 |
| ▶ Bichon frise | 1 |
| ▶ Pomeranian | 1 |
| ▶ Cairn terrier | 1 |
| ▶ Jack Russell (Parson Russell) terrier | 1 |
| ▶ Terrier mix | 1 |
| ▶ Beagle | 1 |
| ▶ Miniature schnauzer | 1 |
| ▶ German shorthaired pointer | 1 |

QuickNotes

When there is evidence of a large IAC and another disease (e.g., granulomatous meningoencephalomyelitis, hydrocephalus) in the same patient, the optimal response to treatment may require treating both conditions.

Diagnosis

Diagnosis of IAC in dogs is typically made via computed tomography (FIGURE 2) or, preferably, MRI.^{2-5,7-10} IACs may also be visualized using ultrasound (via the foramen magnum, temporal window, or persistent bregmatic fontanelle), especially in younger dogs.⁶ The characteristic ultrasonographic appearance of IAC is a large, fluid-filled structure, isoechoic with the CSF spaces (e.g., lateral ventricle) and located between the occipital lobe of the cerebrum and rostral lobe of the cerebellum.⁶

MRI provides the best detail for diagnosis of IAC and is most likely to provide information regarding the presence and nature of concurrent disease states. The typical appearance of IAC on MRI is a well-demarcated, cystic-appearing structure that is hypointense on T1-weighted images, hyperintense on T2-weighted images, and non-contrast enhancing with intravenous gadolinium administration and that suppresses on FLAIR (fluid attenuation inversion recovery) images^{2-5,10,11} (FIGURE 3). Because IAC may be an incidental finding, it is important to rule out concurrent inflammatory disease with a CSF examination. In the absence of an additional brain disorder, the CSF is typically normal in dogs with IAC.

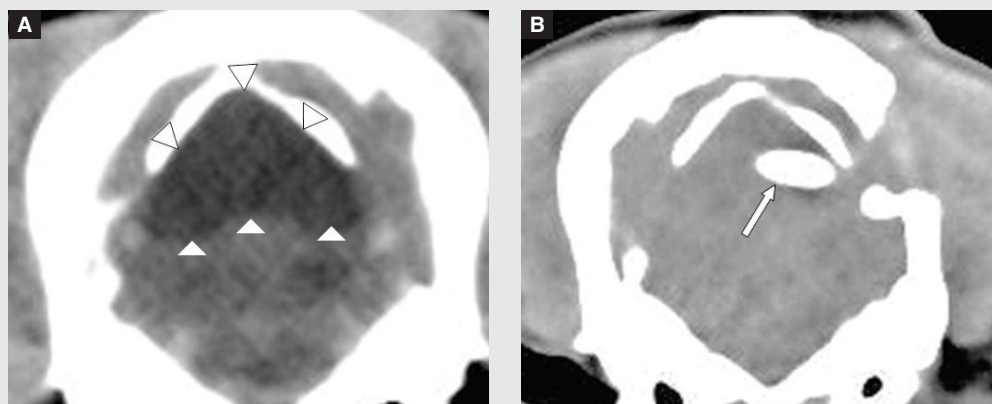
In our opinion, it is often difficult or impossible to discern whether a large IAC in the

presence of another brain disorder is purely an incidental finding. We have seen a number of patients with relatively small dilations of the quadrigeminal cistern (FIGURE 4), which may represent a variant of normal structure or may be evidence of nascent IACs that may be of no clinical significance. Conversely, very large IACs have been described both as sole disease entities and as suspected incidental findings in patients with other intracranial disease processes. Anecdotally, we have also observed similar cystic structures in the brain that do not appear to be associated with the quadrigeminal cistern (FIGURE 5); considering that IACs occur in multiple locations in the human brain, this finding may simply imply that the term *quadrigeminal cyst* is too restrictive in describing IAC in dogs.¹² Since the presence of a large, fluid-filled structure within the cranial vault likely decreases intracranial compliance, some IACs may be contributory rather than incidental findings. In other words, when there is evidence of a large IAC and another disease (e.g., granulomatous meningoencephalomyelitis, hydrocephalus) in the same patient, the optimal response to treatment may require treating both conditions.

In addition, the combined presence of hydrocephalus and IAC in a patient does

FIGURE 2

Transaxial Computed Tomography Images



Preoperative (A) and postoperative (B) transaxial computed tomography images from a dog with an IAC treated with cystoperitoneal shunting. In A, the arrowheads are pointing to the ventral aspect of the cyst and the outlined arrowheads are pointing to the dorsal aspect of the cyst. In B, the arrow is pointing to the rostral aspect of the shunt. (Reprinted with permission from Dewey CW, Krotscheck U, Bailey KS, Marino DJ. Craniotomy with cystoperitoneal shunting for treatment of intracranial arachnoid cysts in dogs. *Vet Surg* 2007;36:416-422.)

not necessarily make one of the disorders (IAC) incidental, nor does this combination ensure that surgically treating one disorder will address the other. Hydrocephalus can be secondary to an IAC, developing because of mechanical obstruction of normal CSF flow by the expanding cyst (i.e., obstructive hydrocephalus).^{10,18,19} It is unlikely that an IAC can be distinguished as communicating or noncommunicating based on standard MRI sequences; such a distinction would likely require either contrast studies or phase-contrast (cine) MRI. In one report of two dogs with IAC, intracystic hemorrhage, which was suspected to have contributed to the development of neurologic dysfunction, was verified at surgery.³

In a recent study, the degree of brain compression by an IAC was calculated from MRI scans of dogs with the disorder, and these measurements were correlated with the presence or absence of clinical dysfunction. It was found that dogs with >14% compression of the occipital lobe always displayed clinical signs and that dogs with compression of both the cerebellum and the occipital lobe were significantly more likely to display clinical signs than dogs with compression of only one region or dogs with no apparent brain compression.¹¹

BOX 2

Clinical Signs Reported in Dogs With Intracranial Arachnoid Cysts*

- ▶ Abnormal mentation
- ▶ Seizures
- ▶ Ataxia
- ▶ Head tilt
- ▶ Strabismus
- ▶ Nystagmus
- ▶ Paraparesis
- ▶ Intention tremors
- ▶ Tetraparesis
- ▶ Visual deficits
- ▶ Neck pain

*Because IAC is often considered an incidental finding, some of these reported clinical signs may be attributable to concurrent intracranial diseases.

Treatment and Prognosis

Because IAC in dogs, as in people, is often considered an incidental finding, it is frequently assumed that treatment of the suspected primary disorder (e.g., inflammatory brain disease, hydrocephalus) is indicated and the IAC is of little or no clinical consequence. As mentioned above, we question whether this approach is appropriate. In cases in which the IAC is considered the primary disease condition, medical therapy is aimed at reducing brain edema and increased intracranial pressure associated with the IAC, as well as controlling seizure activity if present. Medical treatment for IAC is identical to that described for congenital hydrocephalus (e.g., corticosteroids, diuretics, anticonvulsants if indicated), and dose recommendations for various therapies are summarized in **BOX 3**.^{20,21} Dogs with IAC tend to respond initially to medical therapy, but the response may be temporary.

BOX 3

Medical Therapy Options for Dogs With Intracranial Arachnoid Cysts

Glucocorticoids

- ▶ Prednisone: 0.25–0.50 mg/kg PO q12h

Diuretics

- ▶ Furosemide: 0.5–4.0 mg/kg PO q12–24h
- ▶ Acetazolamide: 10 mg/kg PO q6–8h

Proton pump inhibitors

- ▶ Omeprazole: 10 mg/kg PO q24h (dogs <20 kg); 20 mg/kg PO q24h (dogs >20 kg)

Anticonvulsant drugs

- ▶ Phenobarbital: 3–5 mg/kg PO q12h
- ▶ Potassium bromide: 35 mg/kg PO divided q12h
- ▶ Gabapentin: 10 mg/kg PO q8h
- ▶ Felbamate: 15 mg/kg PO q8h
- ▶ Zonisamide: 5 mg/kg PO q12h (if not on phenobarbital; 10 mg/kg if on phenobarbital)
- ▶ Levetiracetam: 20 mg/kg PO q8h
- ▶ Pregabalin: 3 to 4 mg/kg PO q8–12h*

*Dewey CW, Cerda-Gonzalez S, Levine JM, et al. Pregabalin therapy for refractory idiopathic epilepsy in dogs. *J Vet Intern Med* 2008;22:765.

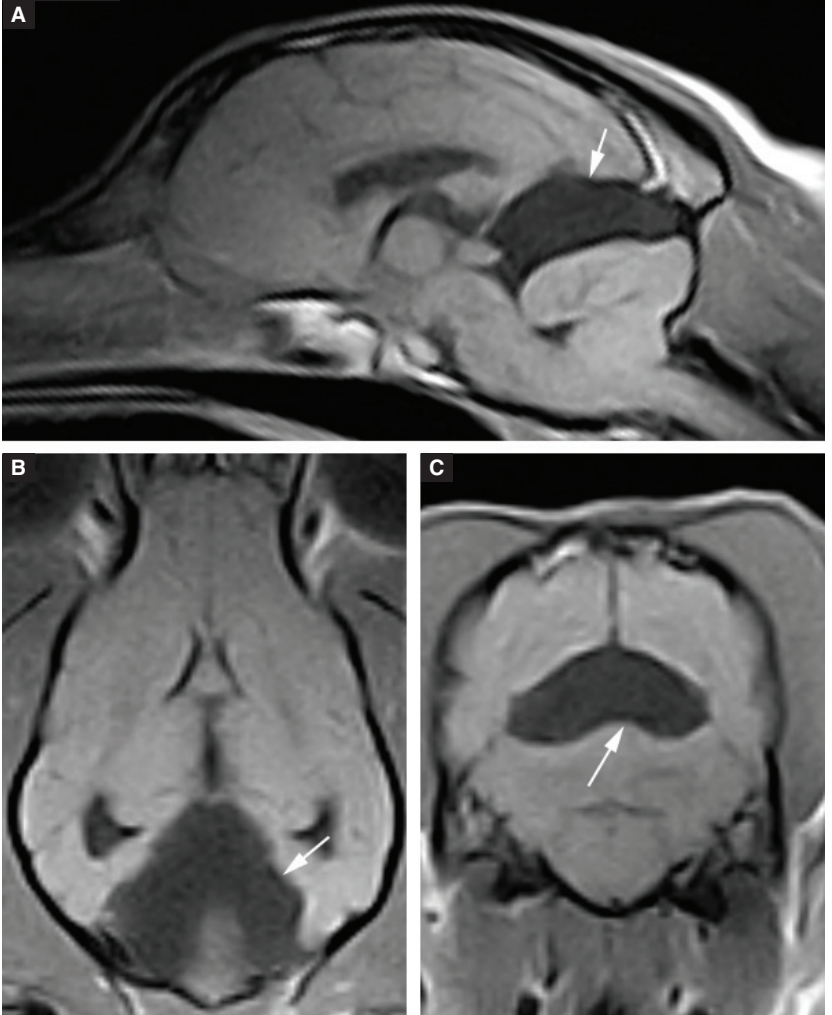
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In cases in which the IAC is considered the primary disease condition, medical therapy is aimed at reducing brain edema and increased intracranial pressure associated with the IAC.

QuickNotes

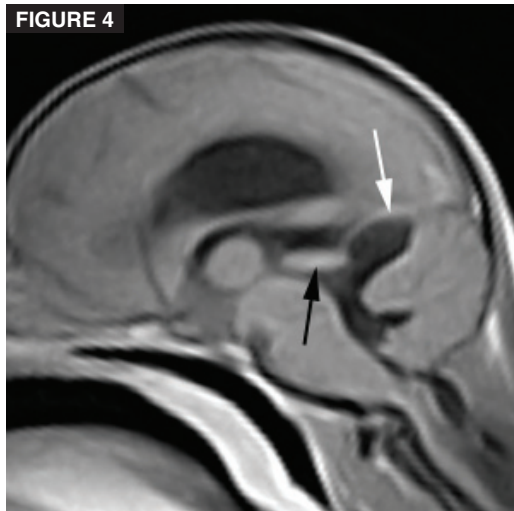
MRI provides the best detail for diagnosis of IAC and is most likely to provide information regarding the presence and nature of concurrent disease states.

FIGURE 3



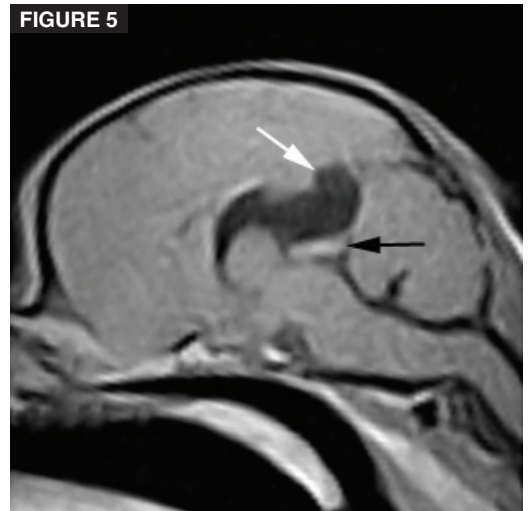
Midsagittal (A), dorsal (B), and transaxial (C) T1-weighted MRI scans demonstrating an IAC in a dog. The arrows are pointing to the cyst in all images.

FIGURE 4



Midsagittal T1-weighted brain MRI scan of a dog with a small dilation of the quadrigeminal cistern (white arrow). The black arrow indicates the dorsal aspect (tectum) of the midbrain.

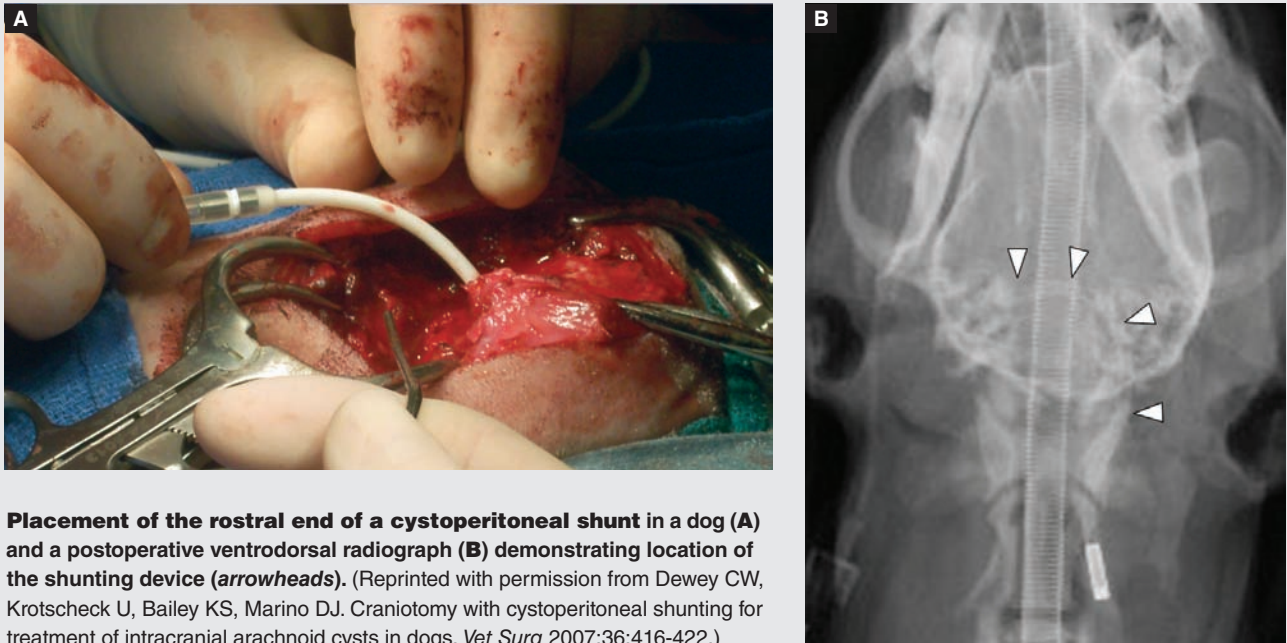
FIGURE 5



Midsagittal T1-weighted brain MRI scan of a dog, demonstrating a cystic structure (white arrow) associated with the third ventricle, rather than the quadrigeminal cistern (black arrow).

FIGURE 6

Cystoperitoneal Shunt Placement



Placement of the rostral end of a cystoperitoneal shunt in a dog (A) and a postoperative ventrodorsal radiograph (B) demonstrating location of the shunting device (arrowheads). (Reprinted with permission from Dewey CW, Krotscheck U, Bailey KS, Marino DJ. Craniotomy with cystoperitoneal shunting for treatment of intracranial arachnoid cysts in dogs. *Vet Surg* 2007;36:416-422.)

Surgical management of IAC in people is typically achieved via either cyst fenestration (i.e., making an opening into the cyst wall) or cystoperitoneal shunt (CPS) placement.^{14,18,19,22-24} Proponents of fenestration cite a high surgical success rate and avoidance of shunt-related complications as reasons for this surgical choice; proponents of CPS report high success rates and avoidance of cyst reexpansion as reasons for the use of this technique.^{13,14,18,19,22-26} Both fenestration and CPS (FIGURE 6) procedures have been reported in dogs with IAC.^{2,3,5,7,10} IAC was considered the primary disease in five reported fenestration cases. Three patients were reimaged after surgery; two of the three dogs had evidence of cyst persistence on MRI. However, only one dog required reoperation.^{2,3,7} Successful CPS of dogs with IAC has also been

reported.¹⁰ The cyst did not reform in any of the shunted cases.

The success rate for surgical management of IAC appears to be high in people and dogs, and whether fenestration or CPS is the preferred procedure remains controversial for both species.¹⁰ Because of the paucity of reports of surgically managed dogs with IAC as well as the suspected high incidence of this abnormality being an incidental finding, the actual surgical success rate for IAC treatment in dogs should be regarded as unknown at this time. Hopefully, as information regarding medical and surgical treatment of clinically significant IAC in dogs accumulates, the understanding of the natural course of this disorder and the effectiveness of medical and surgical therapies to manage it will improve. C

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- In dogs, all reported cases of IAC have been in the _____ fossa.
 - rostral
 - middle
 - caudal
 - none of the above
- Formation of IACs in dogs is believed to be due to
 - a split in the arachnoid meningeal layer during embryogenesis.
 - failure of the neuroectoderm and nonneural ectoderm to separate during embryogenesis.
 - compensatory fluid accumulation following an in utero brain infarction (stroke).
 - failure of the cerebellar vermis to form correctly during embryogenesis.
- Proposed theories to explain progressive expansion of IACs in dogs include
 - active secretion by the arachnoid cells lining the cyst cavity.
 - movement of fluid into the cyst cavity along an osmotic pressure gradient.
 - movement of fluid into the cyst from the neighboring subarachnoid space via slit-like openings (one-way valves) into the cyst lumen.
 - all of the above
- Which is most characteristic of the typical signalment for dog with an IAC?
 - 10-year-old female spayed German shepherd
 - 4-year-old male castrated shih tzu
 - 2-year-old female greyhound
 - There is no typical signalment.
- Clinical signs associated with IAC in dogs include
 - abnormal mentation.
 - generalized seizures.
 - cerebellar dysfunction.
 - all of the above
- The preferred imaging modality for diagnosis of IAC in dogs is
 - ultrasound.
 - scintigraphy.
 - magnetic resonance imaging.
 - computed tomography.
- The characteristic MRI appearance of an IAC in a dog is a large, well-demarcated, cyst-like structure that is
 - hypointense on T1-weighted images.
 - hyperintense on T2-weighted images.
 - contrast-enhancing and hyperintense on FLAIR images.
 - a and b
- In a study of IAC cases in which brain compression by the cyst was measured, dogs were found to be most likely to exhibit clinical dysfunction if
 - the cyst compressed more than 14% of the occipital lobe of the cerebrum.
 - both the occipital lobe of the cerebrum and the cerebellum were compressed by the cyst.
 - a and b
 - none of the above
- Medical therapy for IAC in dogs is directed at
 - decreasing brain edema associated with the cyst.
 - controlling seizure activity if present.
 - minimizing increases in intracranial pressure.
 - all of the above
- Which statement regarding surgical management of IAC is false?
 - Both cyst fenestration and cystoperitoneal shunting procedures have been described in dogs.
 - Cystoperitoneal shunt (CPS) placement has been shown to be superior to fenestration in dogs and humans with IAC.
 - The success rate for surgical management of IAC appears to be high in people and dogs.
 - a and b