



Caudal Occipital Malformation Syndrome in Dogs

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ABSTRACT:

Chiari type I malformation in humans is a congenital abnormality of the caudal occipital bone, resulting in overcrowding of the caudal fossa and compression at the level of the cervicomedullary junction. Both direct bony compression and progressive meningeal hypertrophy at the level of the posterior (dorsal) cervicomedullary junction are believed to lead to abnormalities of cerebrospinal fluid flow dynamics. Central nervous system dysfunction often develops in patients with Chiari type I malformation, with a variety of possible neurologic manifestations. A disorder similar to human Chiari type I malformation occurs in dogs, but characteristic disease features have not been thoroughly described. This article reviews the pathophysiology and clinical features of human Chiari type I malformation and its canine counterpart, caudal occipital malformation syndrome. To determine the characteristic clinical features of the syndrome in dogs, case records of 30 dogs diagnosed with the disorder via magnetic resonance imaging were retrospectively studied. Results of this study suggest that caudal occipital malformation syndrome is relatively common in small-breed dogs and has many clinical similarities to Chiari type I malformation in humans.

Chiari type 1 malformation refers to a developmental abnormality in humans (Figure 1). This disorder is a congenital anomaly of the caudal occipital region of the skull, leading to overcrowding of the caudal fossa and compression of the cervicomedullary junction at the level of the foramen magnum.¹⁻³

Both direct bony compression and progressive meningeal hypertrophy in the region of the posterior (dorsal) cervicomedullary junction are be-

lieved to lead to abnormalities of cerebrospinal fluid (CSF) flow dynamics. In many cases, these CSF pressure changes lead to fluid accumulation within the spinal cord (i.e., syringohydromyelia).^{1,2,4-6} Central nervous system (CNS) dysfunction often develops in humans with Chiari type 1 malformation, with a variety of possible neurologic manifestations. Many of the neurologic manifestations are not associated with secondary syringohydromyelia.^{1,2} A condition that appears to be the canine analog of human Chiari type 1 malformation has recently

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been described in small-breed dogs.⁷⁻¹¹ We propose adopting the more descriptive term *caudal occipital malformation syndrome* to describe the canine disorder. The major difference between human Chiari type 1 malformation and the canine disorder is that many of the reported canine cases have not exhibited obvious cerebellar herniation through the foramen magnum on magnetic resonance imaging (MRI), which is a defining MRI characteristic of Chiari type 1 malformation in humans⁸ (Figure 2).

Most of the literature pertaining to the canine form of the disease is from the United Kingdom, and almost all affected dogs have been cavalier King Charles spaniels. Most reports describe patients with cervical myelopathy due to syringohydromyelia.⁷⁻⁹ Forty cavalier King Charles spaniels from the United Kingdom were recently described with Chiari type 1 malformations; these dogs displayed a wide variety of neurologic presentations, including central vestibular dysfunction,

PATHOPHYSIOLOGY

The cause of caudal occipital malformation syndrome is unknown, but the syndrome is most likely a genetically transmitted developmental disorder of the occipital bone mesoderm.⁸ In humans with Chiari type 1 malformation, there is some evidence of familial aggregation, suggesting a heritable trait.² It was recently proposed that caudal occipital malformation syndrome in cavalier King Charles spaniels is inherited as an autosomal recessive trait, with incomplete penetrance.¹³ It is believed that neuroectodermally derived tissue (i.e., cerebellum, brain stem) in dogs with this syndrome is normal but is crowded into an abnormally small caudal fossa.¹⁻³ In patients with caudal occipital malformation syndrome, there tends to be some level of cerebellar compression as well as constriction of the cervicomedullary junction in the vicinity of the foramen magnum. With chronic bony compression at the cervicomedullary junction and probable turbulent CSF flow

Caudal occipital malformation syndrome in dogs is the canine analog of human Chiari type 1 malformation and appears to be confined to small breeds.

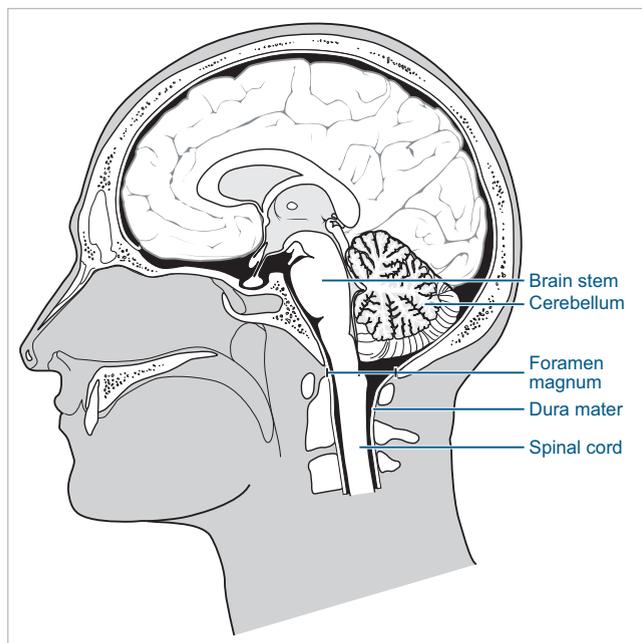
seizure activity, and cervical myelopathy.¹² We have reviewed 30 cases of canine Chiari type 1 malformations diagnosed via MRI at our hospitals from 2001 to 2002. This article reviews the pathophysiology, clinical features, available treatment options, and prognosis for canine Chiari type 1 malformations. Aspects of the human disorder and information from previously published canine cases are discussed. However, the focus is on the 30 recently reviewed cases.

It is our opinion that the presence or absence of cerebellar herniation on MRI in dogs should not be used as a sole discerning feature of the canine disease. Because of the lack of elongated cerebellar tonsils (the portion that typically herniates in humans with Chiari type 1 malformation) in dogs and the horizontal orientation of the canine craniocervical junction (versus vertical in humans), it is less likely for dogs with the same malformation as humans to consistently demonstrate obvious cerebellar herniation on brain imaging. For purposes of discussing disease characteristics, caudal occipital malformation syndrome in dogs is considered the canine analog of Chiari type 1 malformation in humans.

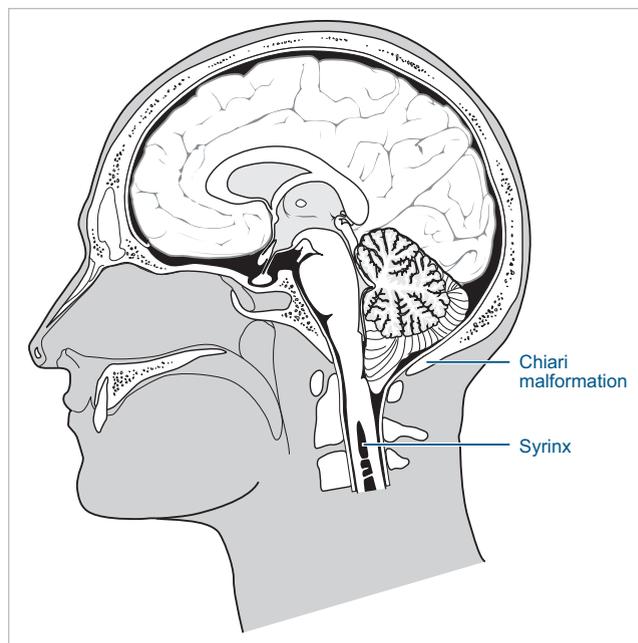
and pressure changes in this region, it is thought that the underlying meninges become hypertrophied with time. In humans, there is pathologic evidence of dural fibrosis in the region of the malformation.⁴ This focal meningeal hypertrophy leads to progression of the constrictive effect at the cervicomedullary junction.

Syringohydromyelia is a common sequela to human Chiari type 1 malformation and typically occurs in the cervical spinal cord region.^{1,2} The same appears to be true in dogs with caudal occipital malformation syndrome. There are numerous theories to explain the development and propagation of syringohydromyelia cavities in patients with caudal occipital malformation syndrome. Common to all the theories is obstruction of normal CSF flow at the level of the cervicomedullary junction (i.e., foramen magnum).^{5-9,11}

In normal dogs, there is pulsatile CSF flow across the foramen magnum from intracranial subarachnoid space to cervical spinal subarachnoid space and back again during systole and diastole, respectively. With an obstruction at the foramen magnum, CSF does not flow well in either direction.^{7,14} However, the pressure exerted

Figure 1. Normal versus malformed occipital/brain-stem region in humans.

Normal configuration of the caudal occipital region in humans.



Chiari malformation in humans. The cerebellum and/or brain stem is located lower than normal and may protrude into the spinal cord. The condition results from a caudal fossa that is too small for the normal cerebellum.

during systole may drive either CSF or a pressure wave from the intracranial compartment into the central canal region of the cranial cervical spinal cord, causing it to progressively expand. This has been referred to as a “water-hammer” effect.^{7,8}

It has also been proposed that CSF is “sucked” into the central canal region of the cervical cord, especially during maneuvers that lead to sudden increases in intrathoracic and intraabdominal pressure (e.g., coughing, sneezing, barking, exercising).^{5,8,15} These Valsalva maneuvers lead secondarily to increased intracranial and intraspinal pressures because of epidural venous distension. Because intracranial pressure is higher than that in the cervical cord region, CSF fluid is drawn into the cervical cord when there are rapid increases in pressure.^{7,8,14,15} Pressure within the spinal compartment tends to increase more rapidly in the lumbar versus cervical regions, further promoting CSF movement into the cervical spinal cord.⁸

The “slosh” phenomenon may also be involved in syringohydromyelia cavity expansion. With distension of epidural veins during events such as coughing or sneezing, CSF flows more freely in the syringohydromyelia cavity than in the compressed subarachnoid space. Therefore, sudden CSF pressure waves cause CSF

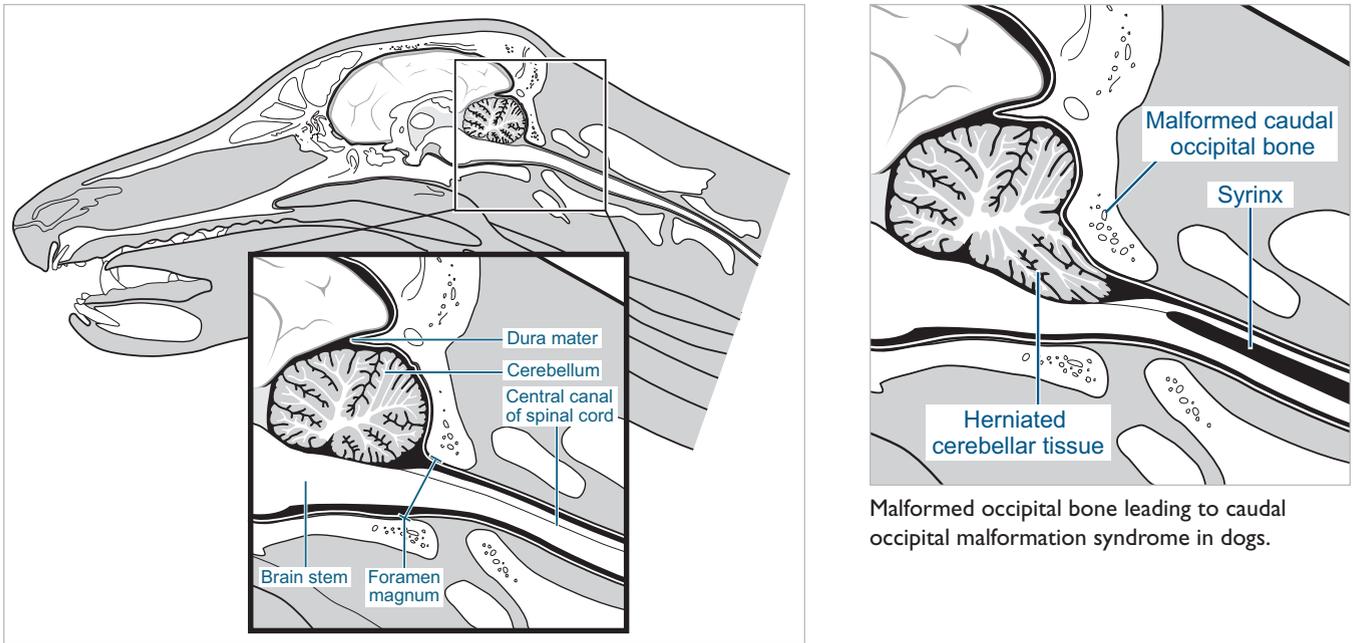
within the cavity to “slosh” around, fissuring surrounding parenchyma and enlarging the fluid cavity.^{5,8,15}

The combination of spinal epidural vein distension (and resultant pressurization of the subarachnoid space) and obstruction to CSF flow from the cervical spine to the intracranial compartment may also result in forcing subarachnoid CSF down perivascular spaces into the spinal cord parenchyma. This leads to progressive enlargement of the syringohydromyelia cavity.^{6,8,15} The possibility of a “ball-valve” effect created by herniated cerebellar tissue at the foramen magnum may add to progressive accumulation of fluid in the cervical cord. If such an effect exists in patients with caudal occipital malformation syndrome, CSF would flow more freely from the intracranial compartment to the cervical region than in the reverse direction.⁷ To what extent these mechanisms actually contribute to the pathogenesis of the syndrome is unknown; however, they all imply that removing the pressure gradient across the foramen magnum should normalize CSF flow in this region.

MRI CHARACTERISTICS

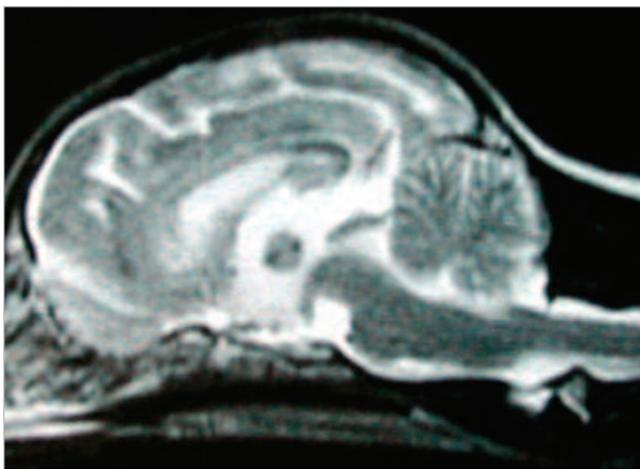
It is unlikely that any imaging modality other than MRI will be used to consistently diagnose caudal occipi-

Figure 2. Normal versus malformed canine occipital/brain-stem region.

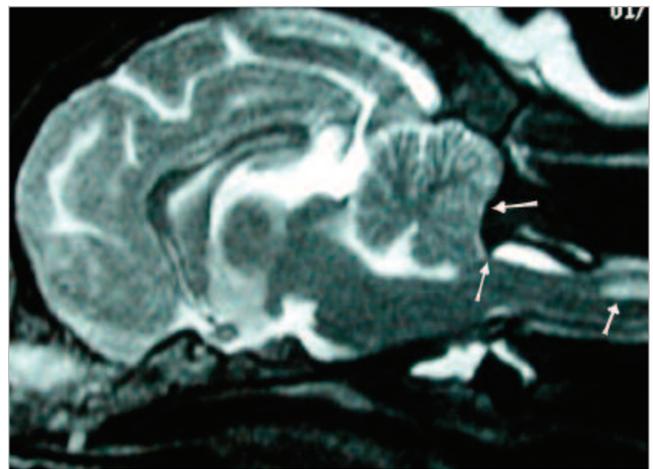


Normal configuration of the canine occipital/brain-stem region.

Figure 3. Midsagittal, T2-weighted MRI brain scans.



A normal dog.

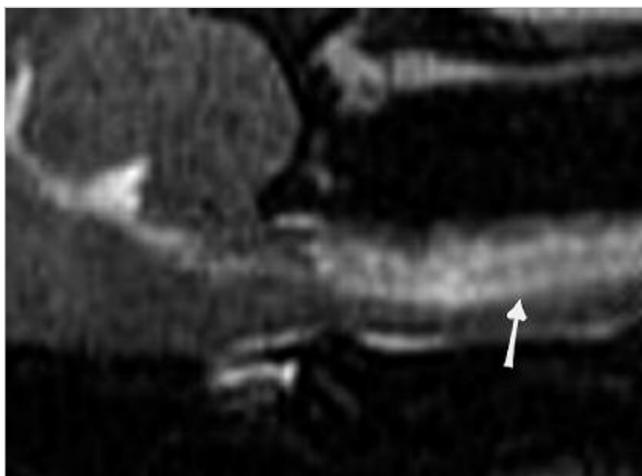


A dog with caudal occipital malformation syndrome. Note the obliteration of the dorsal subarachnoid space at the cervicomedullary junction, rostral displacement of the caudal cerebellum by the occipital bone, and cervical syringohydromyelia cavity (arrows). (From Dewey CW: Myelopathies: Disorders of the spinal cord, in Dewey CW [ed]: *A Practical Guide to Canine and Feline Neurology*. Ames, Iowa State University Press, 2003, pp 277–336; with permission)

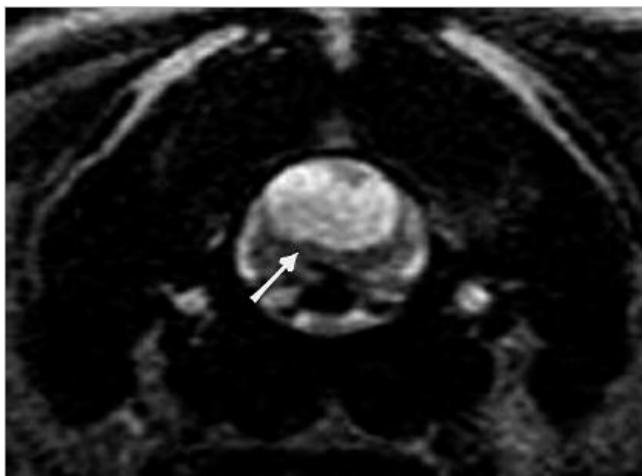
tal malformation syndrome. MRI is also the most effective means of diagnosing syringohydromyelia. The most critical image to evaluate is a midsagittal view (preferably T2-weighted), which includes the caudal fossa and cranial cervical cord^{1,2,6-8,11,14,15} (Figure 3). Characteristic MRI findings in humans with Chiari type 1 malformation include attenuation or obliteration of the posterior subarachnoid space at the level of the foramen magnum (cerebellomedullary cistern), cerebellar tonsillar hernia-

tion through the foramen magnum (at least 5 mm), anterior displacement of the caudal cerebellum by the occiput, and a “kinked” appearance to the medulla.¹⁻³ Other than cerebellar tonsillar herniation, the most con-

Figure 4. T2-weighted MRI demonstrating severe syringohydromyelia of the cervical spinal cord (arrows).



Sagittal view.



Transaxial view.

sistent defining MRI feature of human Chiari type 1 malformation is compression of the CSF space at the level of the cerebellomedullary cistern.^{2,3}

Similar to Chiari type 1 malformation in humans, the most common MRI abnormalities in our series of 30 dogs with caudal occipital malformation syndrome (Table 1) included attenuation or obliteration of the dorsal subarachnoid space at the cervicomedullary junction (all dogs); rostral displacement of the caudal cerebellum by the occipital bone (all dogs); and syringohydromyelia (Figure 4), usually in the cervical cord region (i.e., C2 level caudally). Four (13%) dogs had no MRI evidence of syringohydromyelia. Herniation of the cerebellum through the foramen magnum (Figure 5) was seen in 18

Table 1. MRI Abnormalities in 30 Dogs with Caudal Occipital Malformation Syndrome

Abnormality	Number of Dogs Affected
Attenuation or obliteration of dorsal subarachnoid space at the cervicomedullary junction	30
Rostral displacement of caudal cerebellum by the occiput	30
Syringohydromyelia	26
Cerebellar herniation through the foramen magnum	18
“Kinked” appearance of the caudal medulla	12

(60%) cases. A “kinked” appearance of the caudal medulla (Figure 6) was noted in 12 (40%) dogs. Most (73%) of the dogs had evidence of enlarged lateral ventricles; however, this was considered normal for the breed in all cases. Evidence of other neurologic conditions (e.g., inflammatory foci, disk extrusion/protrusion, intracranial intrarachnoid cysts) was found in 13 (43%) cases.

CLINICAL FEATURES

Chiari type 1 malformation in humans is associated with a wide spectrum of possible neurologic presentations. Clinical signs usually manifest in adulthood, primarily in the second and third decades of life. Pediatric Chiari type 1 malformation is also well described.^{1,2,16} In addition to headache, Chiari type 1 malformation in humans may lead to signs of myelopathy, brain stem dysfunction, and cerebellovestibular dysfunction, either alone or in combination. Scoliosis may occur in cases with syringohydromyelia.^{5,6,14,15} The mechanism of scoliosis development is believed to involve asymmetric loss of paraspinal muscle tone resulting from spinal cord damage caused by the syringohydromyelia cavity. The muscle tone loss may be due to dysfunction of lower motor neurons (LMNs) that innervate these muscles, interference with sensory input to the LMNs, or a combination of these phenomena.^{8,14,17} Seizures reportedly occur in approximately 10% to 12% of humans with Chiari type 1 malformation. Although the mechanism for seizures with this disorder is not certain, there is some evidence that the normally functioning cerebellum

Table 2. Clinical Manifestations of Caudal Occipital Malformation Syndrome in 30 Dogs

Clinical Manifestation	Number of Dogs Affected
Cervical or cranial hyperesthesia	12
Multifocal CNS dysfunction	9
Cervical myelopathy	9
Cerebellovestibular dysfunction	5
Forebrain dysfunction (including seizure activity)	5
Persistent scratching at shoulder region	2
Torticollis	3
Bilateral cranial nerve VII paralysis	1

has an inhibitory effect on seizure activity. Other theories proposed to explain seizure activity in association with Chiari type 1 malformation include concurrent cerebral dysgenesis or hydrocephalus.^{18,19} Asymptomatic Chiari type 1 malformation (with or without syringohydromyelia) has also been described^{20,21}; in one report, 14% of patients were found to be asymptomatic.²⁰ Myelopathy in humans with Chiari type 1 malformation is not always associated with syringohydromyelia. In one study, 35% of humans with symptomatic Chiari type 1 malformation did not have evidence of syringohydromyelia; however, 66% of them exhibited evidence of myelopathy.²

The literature concerning canine caudal occipital malformation syndrome has focused on patients with cervical myelopathy secondary to syringohydromyelia.^{7-11,14} These dogs typically exhibited thoracic limb paresis (often LMN in nature) that was worse than pelvic limb paresis, a phenomenon referred to as *central cord syndrome*.^{8,14} Cervical spinal hyperesthesia and persistent scratching at the neck and shoulder regions have also been described. It is generally believed that the scratching represents paresthesia secondary to syringohydromyelia fluid interfering with spinothalamic tracts and/or dorsal horn neurons.^{8,9}

The mean age of dogs in our caudal occipital malformation syndrome group was 6.26 years of age (range: 4.5 months to 13 years of age). There were 17 males (two



Figure 5. Midsagittal, T2-weighted MRI of the cervicomedullary junction in a dog with caudal occipital malformation syndrome demonstrating cerebellar herniation through the foramen magnum (arrow).

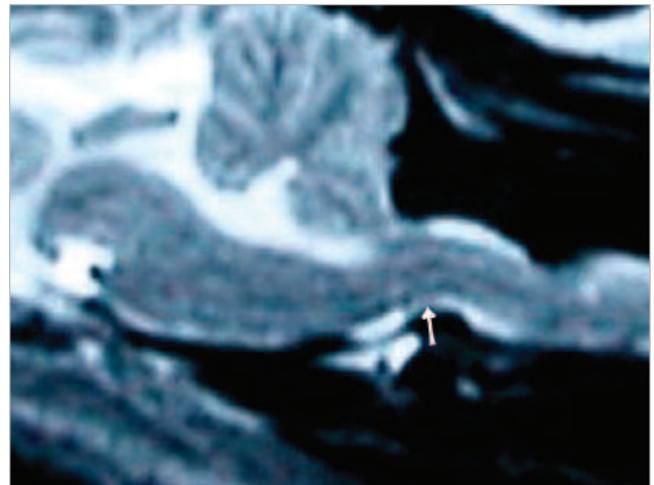


Figure 6. Midsagittal, T2-weighted MRI of a dog with caudal occipital malformation syndrome demonstrating a "kinked" appearance of the medulla oblongata (arrow).

intact) and 13 females (two intact). All were small-breed dogs, with Yorkshire terriers (seven) and miniature or toy poodles (six) being the most common. Other breeds included bichon frise (two), pug (two), Chihuahua (two), West Highland white terrier (two), cavalier King Charles spaniel (two), shih tzu (two), Maltese (one), miniature

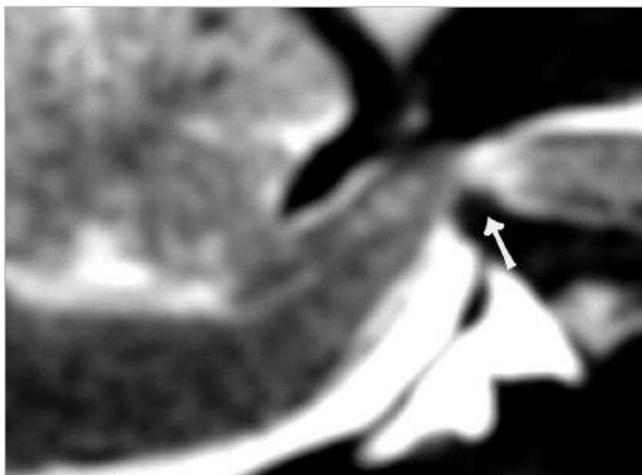


Figure 7. Midsagittal, T2-weighted MRI of a dog with caudal occipital malformation syndrome demonstrating dorsal angulation of the dens (arrow).

pinscher (one), Pomeranian (one), Pekingese (one), and French bulldog (one). One of the cavalier King Charles spaniels was known to have been acquired from Ireland. As with human Chiari type 1 malformation, there was a wide variety of neurologic presentations in the dogs (Table 2). Nine dogs had evidence of multifocal CNS dysfunction. Clinical signs of myelopathy were common (19 [63%]) but were a component of multifocal CNS dysfunction in eight of the cases. Isolated cervical myelopathy occurred in nine (30%) dogs. Cerebellovestibular dysfunction was also prevalent (nine [30%]), either alone (five [17%]) or as a component of

One dog was felt to have incidental (asymptomatic) caudal occipital malformation syndrome. The dog had a history of poorly localizable pain somewhere in the spine. After MRI of the entire spine, a large L7-S1 disk protrusion was found, along with mild deformation of the caudal cerebellum. This dog's clinical signs resolved completely after the cauda equina region had been decompressed. Five dogs had evidence of disk extrusion/protrusion, most of which was subjectively judged to be mild. Four dogs had evidence of inflammatory brain disease based on parenchymal lesions and CSF results. Two of these were suspected to have Yorkshire terrier encephalitis, one was suspected to have granulomatous meningoencephalomyelitis, and one had lesions consistent with CNS lymphosarcoma at necropsy. Two dogs had evidence of an intracranial intraarachnoid cyst (quadrigeminal cyst) in the caudal fossa. On MRI, one dog had severe dorsal dens angulation (Figure 7), a condition recently reported in conjunction with caudal occipital malformation syndrome in a dog.¹⁰

Excluding the four dogs with suspected concurrent inflammatory brain disease, CSF evaluation was available for 12 dogs. CSF results were normal in nine of these dogs. Two had evidence of mild mononuclear pleocytosis (10 and 20 leukocytes/ μ l, respectively; normal: <5 cells/ μ l), and one had a normal cell count with an elevated protein level (66.2 mg/dl; normal: <48 mg/dl).

TREATMENT

Follow-up information was available for 29 of 30 patients. Most of the symptomatic dogs (25 of 28) were

Caudal occipital malformation syndrome in dogs is characterized by a wide range of clinical presentations, including cerebellovestibular dysfunction, cervical myelopathy, and seizure activity.

multifocal CNS disease (four [13%]). Five (17%) dogs, all of which had seizure activity, exhibited isolated fore-brain dysfunction. Twelve (40%) demonstrated evidence of cervical and/or cranial hyperesthesia during palpation. Persistent scratching at the shoulder region occurred in two dogs. Torticollis was observed in three dogs. Dogs with scratching and torticollis had evidence of syringomyelia in the cervical spine. One dog exhibited bilateral cranial nerve VII paralysis.

treated with antiinflammatory doses of oral glucocorticoids (e.g., prednisone [0.5 mg/kg q12h]). Additional therapies in three of these dogs included chlorambucil (in a dog with suspected necrotizing encephalitis), methazolamide (in a dog with multiple intervertebral disk extrusions/protrusions), and potassium bromide (in a dog with caudal occipital malformation syndrome alone and seizures). One dog with seizures as the sole clinical sign of dysfunction was treated with phenobarbital

alone. Two symptomatic patients received no therapy. One of these dogs had cervical myelopathy and evidence of cervical intervertebral disk extrusions/protrusions on MRI. The other dog showed MRI evidence of caudal occipital malformation syndrome alone and had exhibited clinical signs of both cerebellovestibular and cervical spinal cord dysfunction. One dog had several large cervical disk extrusions/protrusions evident on MRI in addition to a quadrigeminal cyst. The patient was non-ambulatory tetraparetic and underwent dorsal cervical laminectomy following poor response to prednisone therapy. Five dogs that had initially been treated with glucocorticoids were treated surgically via foramen magnum decompression (Figure 8). In all of these dogs, the only disease process identified was caudal occipital malformation syndrome. In four of five of these dogs, meningeal resection (i.e., dura, arachnoid) over the region of decompression was also performed. Meningeal tissue from two dogs was submitted for histopathologic evaluation and was determined to be dural fibrosis.

The treatment of choice for humans with Chiari type 1 malformation is surgical decompression of the foramen magnum.^{1,8,22-24} Although numerous surgical procedures have reportedly been used in treating the human disorder (e.g., syringohydromyelia cavity shunting, cerebellar tonsil resection, obex plugging), most recent reports advocate foramen magnum decompression.²²⁻²⁴

PROGNOSIS

The prognosis for humans with Chiari type 1 malformations treated with foramen magnum decompression is often favorable, especially when surgery is performed early in the disease process. Most patients experience either halted progression of signs or sustained improvement of clinical signs.²²⁻²⁴

When this article was submitted for publication, nine of 29 (31%) patients available for follow-up had died (one) or been euthanized (eight) because of neurologic disease. Five of these dogs had evidence of caudal occipital malformation syndrome alone, and four had evidence of concurrent neurologic disorders. One of the deceased dogs died during surgery (dorsal decompression for multiple large cervical intervertebral disk extrusions/protrusions) after a 2-week course of prednisone failed to improve its neurologic function. Three other dogs were euthanized within 1 to 4 days of diagnosis. One of these dogs had lymphocytic pleocytosis (suspected lymphoma) and had improved slightly after prednisone administration, one dog had a quadrigeminal cyst, and one dog had

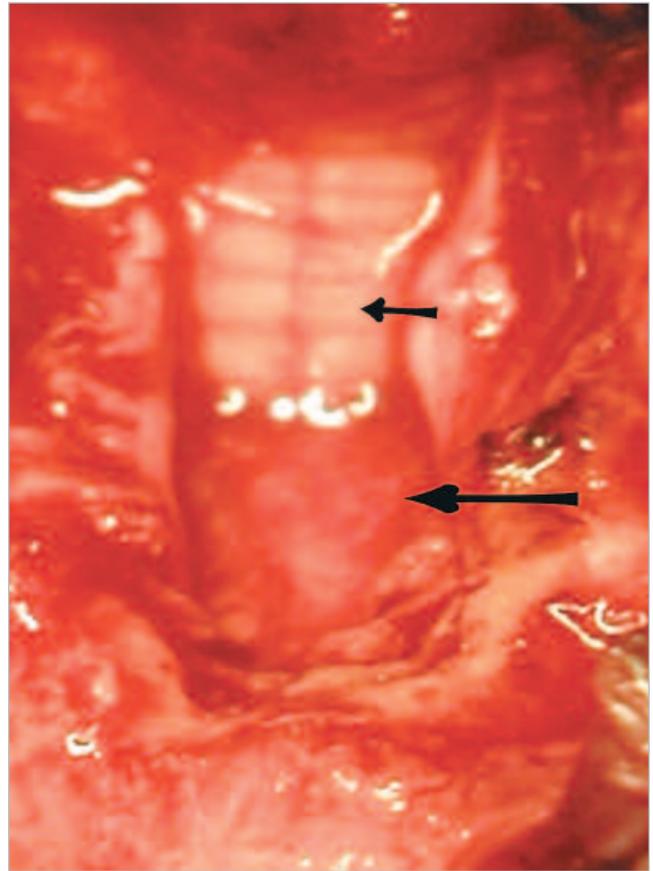


Figure 8. Intraoperative dorsal view of foramen magnum decompression performed on a dog with caudal occipital malformation syndrome. The meninges have been removed, exposing the cerebellum (short arrow) and medulla oblongata (long arrow).

caudal occipital malformation syndrome alone. The dog with the cyst worsened despite prednisone therapy, and the dog with caudal occipital malformation syndrome alone did not improve with prednisone therapy. Follow-up times for the remaining 25 dogs ranged from 3 to 128 weeks (mean: 45.4 weeks). Of these 25 cases, clinical signs of dysfunction resolved in five (20%), improved in eight (32%), were unchanged in five (20%), and worsened in seven (28%).

Three of five patients that experienced clinical resolution were dogs with caudal occipital malformation syndrome alone that had undergone foramen magnum decompression surgery. The other two cases that resolved included the dog with dorsal dens angulation and the one with presumed asymptomatic caudal occipital malformation syndrome and lumbosacral disk disease. Clinical signs of dysfunction improved in two of

the remaining five dogs treated with foramen magnum decompression. One of these dogs improved dramatically but retained torticollis. This dog did not undergo meningeal resection. The other dog that improved but was not cured after foramen magnum decompression had been nonambulatory paraparetic; when this article was submitted for publication, this dog was ambulatory and had pelvic limb monoparesis. Glucocorticoids were tapered and eventually discontinued in the five dogs treated with foramen magnum decompression. Clinical signs improved in two dogs with caudal occipital malformation syndrome alone that were treated with foramen magnum decompression as well as in six other dogs, including three with caudal occipital malformation syndrome alone, two with suspected necrotizing encephalitis, and one with multiple intervertebral disk extrusions/protrusions.

Two of five dogs that experienced no change in clinical status were not treated. One dog had caudal occipital malformation syndrome alone, and the other had multiple intervertebral disk extrusions/protrusions. Among

The disorder appears to be limited to small-breed dogs. In our group of 30 patients, multiple breeds were represented, and only two were cavalier King Charles spaniels. Both males and females are affected, and there seems to be a wide age range at the onset of clinical signs. Diagnosis of caudal occipital malformation syndrome largely depends on MRI, especially a midsagittal view of the caudal brain and cranial cervical spinal cord. Consistent MRI abnormalities supporting diagnosis of the syndrome are attenuation or obliteration of the dorsal subarachnoid space at the cervicomedullary junction (cerebellomedullary cistern) and rostral displacement of the caudal cerebellum by the occiput. A large proportion (43%) of the cases had concurrent neurologic disorders. To what extent these disorders contributed to the disease process was unclear. In some cases, it is possible that either caudal occipital malformation syndrome or the concurrent neurologic condition is incidental. It is also possible that the syndrome and other conditions that increase pressure within the spinal compartment (e.g., disk extrusions) can exacerbate each other. Dogs

Treating caudal occipital malformation syndrome and its sequelae may include medical therapy and/or surgery.

the other three patients, one had caudal occipital malformation syndrome alone, one had multiple disk extrusions/protrusions, and one had multiple disk extrusions/protrusions and middle ear disease.

Five of seven dogs that worsened were euthanized. Four of these dogs had caudal occipital malformation syndrome alone, and one was suspected of having granulomatous meningoencephalomyelitis. All five dogs initially responded favorably to glucocorticoid therapy and subsequently worsened; two of five dogs worsened after prednisone was tapered (because of unacceptable side effects). One of two surviving dogs that worsened had caudal occipital malformation syndrome alone; this dog's sole clinical abnormality was seizure activity, which increased in frequency despite phenobarbital therapy. The remaining dog that worsened had multiple disk extrusions/protrusions.

SUMMARY

Like humans with Chiari type 1 malformation, dogs with caudal occipital malformation syndrome can present with a wide variety of neurologic abnormalities.

with caudal occipital malformation syndrome may be more sensitive to the effects of diseases that increase intracranial or intraspinal pressure. Because CSF pressure within the caudal fossa is believed to increase with caudal occipital malformation syndrome, the quadrigeminal cysts noted in two of 12 dogs with concurrent abnormalities may be a result of the syndrome rather than an incidental finding.

Like other reports, this investigation suggests that most dogs with caudal occipital malformation syndrome improve when treated with glucocorticoids, but clinical signs of dysfunction do not resolve. Five surgical cases do not provide enough evidence to make a valid judgment; however, the apparently positive response of these five dogs to foramen magnum decompression suggests that surgery may play an important role in managing the syndrome, as does the same procedure in managing human Chiari type 1 malformation.

It is likely that canine caudal occipital malformation syndrome is not a rare disorder. Rather, increased awareness of this syndrome in dogs is probably due to increased availability of MRI for veterinary patients.

Resources

Web (human sites)

The Chiari Institute
www.chiariinstitute.com

Columbia-Presbyterian
Chiari Malformation
http://cpmcnet.columbia.edu/dept/nsg/PNS/
ChiariMalformation.html

Duke Center for Human Genetics
Chiari Malformations and Syringomyelia Fact Sheet
www.chg.duke.edu/patients/facts.html

University of Missouri
The Chiari Clinic
http://tribble.missouri.edu/ns/chiari/

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The syndrome should be considered in the differential diagnosis of small-breed dogs with CNS dysfunction, especially if diagnostic test results (e.g., myelography, computed tomography, CSF evaluation) have not been productive.

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ARTICLE #5 CE TEST



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I. Caudal occipital malformation syndrome in dogs is believed to be primarily due to

- a. congenital malformation of the brain stem and cerebellum.
- b. a congenital abnormality of the occipital bone, with secondary overcrowding of the caudal fossa.
- c. an infectious agent that leads to multifocal CNS dysfunction.
- d. secondary compression of the brain stem by a primary syringohydromyelia cavity in the cervical spinal cord.

- 2. Which effect may help explain progressive pressure abnormalities in the spinal compartment and subsequent development of syringomyelia?**
- “water-hammer”
 - “suck”
 - “slosh”
 - all of the above
- 3. The most important test in diagnosing caudal occipital malformation syndrome in dogs is**
- CSF examination.
 - myelographic evaluation of the cervical spinal cord.
 - computed tomography of the brain stem and cerebellum.
 - midsagittal MRI of the cervicomedullary junction.
- 4. The most consistent MRI abnormality in cases of caudal occipital malformation syndrome is**
- attenuation or obliteration of the dorsal subarachnoid space at the cervicomedullary junction.
 - rostral displacement of the caudal cerebellum by the occipital bone.
 - herniation of the caudal cerebellum through the foramen magnum.
 - a and b
- 5. Concurrent neurologic disorders _____ dogs with caudal occipital malformation syndrome.**
- rarely occur in
 - are apparent in all
 - have been identified in over 40% of
 - have been identified in less than 10% of
- 6. Clinical features of caudal occipital malformation syndrome in dogs include**
- cervical myelopathy.
 - cerebellovestibular dysfunction.
 - multifocal CNS dysfunction.
 - all of the above
- 7. Dogs with caudal occipital malformation syndrome have reportedly been**
- exclusively small breeds.
 - almost exclusively cavalier King Charles spaniels in Europe.
 - exclusively Yorkshire terriers in the United States.
 - a and b
- 8. The preferred treatment for symptomatic Chiari type I malformation in humans is**
- foramen magnum decompression.
 - high-dose glucocorticoid therapy.
 - ventriculoperitoneal shunt placement for concurrent hydrocephalus.
 - oral administration of diuretic drugs.
- 9. In dogs with caudal occipital malformation syndrome that are treated with glucocorticoid therapy alone,**
- clinical signs tend to resolve completely.
 - clinical signs tend to remain unchanged.
 - clinical signs tend to improve but do not often resolve.
 - side effects of therapy do not occur.
- 10. In treating caudal occipital malformation syndrome in dogs, foramen magnum decompression**
- has little promise because it typically fails to improve the neurologic status of humans with Chiari type I malformation.
 - may be useful but should probably include meningeal resection.
 - is not viable because most dogs can be treated with glucocorticoid therapy alone.
 - may be useful, but meningeal resection is strictly contraindicated in dogs.