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Developmental neurological disorders of the dog and cat

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INTRODUCTION

Many developmental neurological disorders result in death in utero or shortly after birth. Others may be slowly progressive and clinical expression may be delayed. From an imaging perspective, developmental neurological disorders can be separated into those of primary neural origin versus those of skeletal origin that secondarily affect the neurological system. Developmental disorders of primary neural origin may further be differentiated into those that result in a gross anatomical abnormality and those that have an underlying metabolic cause that affect the neurological system at the subgross or microscopic level.

Skeletal Anomalies that Clinically Affect the Central Nervous System

Vertebral Anomalies

Complex thoracic vertebral anomalies: Thoracic vertebral anomalies are common in English Bulldogs, Pugs, Boston Terriers and other brachycephalic breeds and most often involve the mid thoracic spine. Complex spinal malformation involving hemivertebrae, butterfly vertebrae, block vertebrae result from abnormal development, ossification and fusion of the vertebral ossification centers. These malformations often lead to some combination of scoliosis, kyphosis, lordosis and spinal canal stenosis. Although alterations in the spinal canal may prevent normal spinal cord development, adaptation may initially minimize the extent of neurological compromise. Because the stability of the spine may also be affected, patients with complex vertebral anomalies often develop clinically appreciable neurologic deficits as a result of what might be considered relatively minor trauma.

Spina Bifida: This entity results from a failure of fusion of the dorsal ossification centers during development and most commonly involves the caudal lumbar and sacral/coccygeal vertebral. This anomaly may be restricted to the vertebral but may also incorporate the meninges (meningocele) or the meninges and spinal cord (meningomyelocele). This disorder may be part of the sacrococcygeal dysgenesis seen most commonly in English Bulldogs and Manx cats.

Cervical Spondylomyelopathy: This is a somewhat poorly defined disorder of the cervical spine that secondarily affects the cervical spinal cord. There are two general presentations. The first tends to occur in middle age Doberman pinschers and other large breed dogs and clinically manifests as ventral compression of the caudal cervical spine due to some combination of vertebral subluxation, ligamentous hypertrophy and intervertebral disk protrusion. The second form, more germane to this discussion, occurs in young, large breed dogs and appears as a dorsal and lateral compression of the cervical spinal cord due to articular facet productive remodeling in conjunction with synovial and ligamentous hypertrophy and stenosis of the spinal canal.

Atlanto-Axial Anomalies: Cervical instability due to atlanto-axial anomalies is most common in small and toy breed dogs. A range of anomalies may be present but malarticulation between the two vertebral segments is the common element. This disorder often includes aplasia or hypoplasia of the odontoïd process. Clinical manifestations may be limited to cervical pain on manipulation but often include neurological deficits as the result of dynamic spinal cord compression. Depending on the degree of instability, the disorder may be recognized only after a minor traumatic incident.

Skull Anomalies

Chiari-like Malformation: This is a complex anomaly involving the caudal fossa of the skull, the cerebellum and the cervical spinal cord. The disorder is most common in Cavalier King Charles Spaniels and is thought to be caused, in large part, by underdevelopment of the caudal fossa. Diagnosis is made through a combination of signalment, clinical signs and imaging findings. Magnetic resonance imaging features include, occipital hypoplasia, caudal cerebellar herniation through the foramen magnum and cervical syringomyelia.

INTRACRANIAL DEVELOPMENTAL DISORDERS

Developmental Disorders With Anatomical Presentation

Caudal Fossa / Cerebellar Anomalies

Intracranial Arachnoid Cysts: These lesions most commonly occur in toy breed dogs and most involve the quadrigeminal cistern. Arachnoid cysts may be associated with seizures and ataxia or may be clinically silent. Arachnoid cysts are easily identified as well-delineated fluid-filled intracranial masses causing distortion and displacement of the cerebellum when large.
Cerebellar Disorders: Clinical signs for these disorders may be similar, all reflecting diminished cerebellar function including poor motor function, tremors and uncoordinated movement. MRI is used to document alterations in normal cerebellar anatomy which may appear as reduced cerebellar volume and increased prominence of the adjacent subarachnoid space with cerebellar atrophy and hypoplasia and be aplasia or hypoplasia of the cerebellar vermis and enlargement of the fourth ventricle in Dandy-Walker syndrome.

Cerebellar Aplasia: This is an exceedingly rare usually autosomal recessive disease caused by death of Purkinje cells in the cerebellum beginning shortly after birth.

Cerebellar Hypoplasia: This is another rare disorder of cats and dogs that is thought to be induced by exposure to a variety of infectious agents (most commonly feline panleukopenia virus in cats) while in utero.

Dandy–Walker Syndrome: Is yet another rare disorder characterized by hypoplasia or aplasia of the cerebellar vermis and 4th ventriculomegaly.

Rostral Fossa

Congenital Hydrocephalus: This disorder is usually caused by disruption of CSF production and resorption (usually from obstruction) in utero leading to distension of the ventricular system. In advanced cases, the cortical mantle may be very thin. The disorder is easily detected on MR examination.

Lissencephaly: This is a rare disorder in which neuronal migration is disrupted in utero leading to a lack of formation of the normal cortical gyri and sulci (agyria or pachygyria). The disorder is best confirmed on T2-weighted MR images which reveals a smooth or mildly undulating cortical surface.

Polymicrogyria/Schizencephaly: There are a number of developmental brain disorders due to abnormal neuronal migration that result in abnormal appearance of the brain as well as large abnormal accumulations of CSF. MRI is used to characterize the brain parenchymal abnormalities as well as defining distribution of CSF.

Disorders of the Brain with Metabolic or Idiopathic Causes

Lysosomal Storage Disorders: This term includes a wide variety of predominantly autosomally recessively inherited metabolic disorders. Specific enzyme deficiencies result in improper metabolism of lipids, glycoproteins or mucopolysaccharides that, in turn, adversely affect neurons, myelin or glial cells. Clinical presentation and age of onset of neurologic signs varies depending on the specific disorder but usually occurs by 11 months of age. Some of these disorders cause multisystem disease. The MR imaging appearance of these disorders are not well reported and vary widely but may include cortical atrophy, loss of definition between gray and white matter and diffuse or multifocal intensity changes of brain parenchyma.

Husky Encephalopathy: A number of isolated or rare inherited disorders affecting specific sites in the brain have been reported. Husky Encephalopathy is an example of such a disorder that has been well described. Dogs developed clinical signs usually within 11 months of age that included ataxia, visual deficits, compulsive behavior abnormal prehen-

Toxicities

Some neurotoxicities, such as hepatoencephalopathy, may sometimes occur due to developmental anomalies involving other organ systems (portosystemic shunt). A number of other exogenous toxic insults to the central nervous system may occur in young animals and clinical and imaging findings can mimic those of developmental disorders. Two illustrative examples are included below.

Hepatoencephalopathy: Intracranial neurological deficits frequently occur as a sequelum to portosystemic shunts anomalies as a result of increased circulating concentrations of ammonia and other related metabolites. Typical signs range from subtle reductions in cognitive function to obtundation and coma. MR imaging characteristics have been reported to include prominence of the sulci suggesting cortical atrophy and focal hyperintensity of the lentiform nuclei on T1-weighted images that do not contrast enhance. In our experience, MR features are often more widespread and include diffuse T2 hyperintensity presumably due to increased permeability.

Thiamine Deficiency: Patients are presented with nonspecific clinical signs of inappetence, weight-loss, and multifocal intracranial disease. MR features include hyperintensity of the caudate nucleus and rostral colliculi on T2 weighted images.

Spinal Cord

Spinal Dysraphism: A number of congenital anomalies involve incomplete fusion of the dorsal elements of the spine (Spinal dysraphism). Some include simple incomplete fusion of the dorsal aspect of the cord while others result in masses consisting of meningeal distension (meningocele) or meningeal distension with neural elements incorporated (myelomeningocele). These anomalies may result in the tethered cord syndrome often presenting clinically with excretory incontinence.

Hydromyelia/Syringomyelia: these terms refer to pathologic dilatations of the central canal (hydromyelia) or within spinal cord parenchyma (syringomyelia). These may be primary entities or may be secondary to other anomalies. (e.g. cervical hydromyelia associated with Chiari-like syndrome of Cavalier King Charles Spaniels). Often the two entities cannot be clearly distinguished on MR images but both result in focal or regional, usually elongate, accumulations of CSF that appears hypointense on T1 images and hyperintense on T2 images.

Arachnoid Cysts: This disorder most commonly involves the cranial cervical in young dogs. Focal dilatation of the subarchnoid space (usually dorsally located) often occurs in concert with focal atalation in the diameter and shape of the spinal cord at the same level. Neurologic deficits depend on the location and size of the cyst and corresponding cord changes.
**Dermoid sinus:** This rare anomaly results in incomplete separation of spinal and skin elements. Sinuses may form a blind sac or may extend to the subarchanoid space. Fistulography, myelography, MRI and contrast enhanced CT may all be used for evaluating the extent of the lesion.

**Additional reading**