Spina Bifida, Meningomyelocele, and Meningocele

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KEYWORDS

• Congenital malformation • Meningocele • Meningomyelocele • Neural tube defect • Spina bifida

KEY POINTS

- The cause of spina bifida with or without meningomyelocele or meningocele (MC) is likely related to genetic and environmental factors.
- Spina bifida with or without meningomyelocele or MC can be associated with other congenital abnormalities of the vertebrae, central nervous system (CNS), and adjacent soft tissues.
- A thorough physical and neurologic examination is warranted in all animals in which an anomaly is suspected.
- Consideration should be given to imaging the entire CNS and adjacent structures to identify coexistent malformations.
- Surgical interventions may be considered in select animals to potentially prevent worsening of signs. In some cases, neurologic improvement may also occur.

INTRODUCTION

Neural tube defects (NTD) are a collection of congenital malformations that typically occur as a result of abnormal development and/or closure of the neural tube during embryogenesis. Development of the central nervous system (CNS) is a precisely timed and highly regulated process that is integrated intimately with the development of the vertebrae, paravertebral musculature, and overlying skin, in addition to other structures such as the distal gastrointestinal and urinary tracts. Consequently, whereas the term NTD connotes anomalous development of the CNS, more broadly, NTDs

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encompass anomalous development of the vertebrae, paravertebral muscles, and skin in addition to the CNS, and implies the possible coexistence of anomalous development of other anatomic structures. NTDs can be further classified based on whether they are open, exposed neural tissues and leaking cerebrospinal fluid (CSF), or closed, not exposed neural tissues and not leaking CSF.

The terminology used in the literature to describe NTDs is often nonspecific, confusing, or inconsistently applied. **Table 1** contains common terms and their definitions. In particular, several terms are used synonymously with NTD, including spina bifida and spinal dysraphism. Spina bifida is the most commonly used term to describe NTD. Strictly speaking, spina bifida is the embryologic failure of fusion of 1 or more vertebral arches (laminae); subtypes of spina bifida are based on the degree and pattern of malformation associated with neuroectoderm involvement.[1] It can be qualified as aperta (open), cystica (closed), or occulta (hidden or have a concealed external demarcation). Spina bifida alone may not be associated with clinical signs; however, it may be accompanied with spinal cord malformation. The Greek origin of the term *raphe* means the line of union of 2 contiguous bilaterally symmetric structures.[1] In the context of the neural tube, there is an implication of a lack of fusion along the midline of the neural tube.[1] Although terms such as spina bifida and dysraphism are often used in the literature interchangeably, the more broad term NTD may be preferable. Where possible, the authors have tried to use the term NTD to not only encompass spinal cord malformations and vertebral malformations, but also to focus attention on a common pathophysiologic basis notable for neural tube development.

The most common NTDs in humans include spina bifida and anencephaly.[11,12] The cause of NTDs is multifaceted, with contributions from a varying combination of genetic predispositions and environmental interactions. Human estimates for the prevalence of spina bifida in the United States are currently 3.39 per 10,000 live births.[11,13] NTDs in domestic animals are likely underreported with reports limited to single cases and small case series. Although the current prevalence in dogs and cats is unknown, previous reports suggest a prevalence of spina bifida of 0.006% in dogs and 0.09% in cats.[14]

NTDs can occur anywhere along the vertebral column although the majority of reported spina bifida and associated meningomyelocele (MMC) or meningocele (MC) cases occur within the lumbosacral spinal cord and vertebral column. Animals with lumbosacral NTDs display characteristic physical, neurologic, and diagnostic findings. Early recognition of NTDs may allow for improved outcomes for affected animals. The goal of this article is to provide a summary of the embryology and the proposed mechanisms for the congenital defects, outline the clinical signs, diagnostic findings and treatment options that are available for dogs and cats today.

EMBRYOLOGY

The development of the CNS is initiated with the formation of the neuroectoderm, which is derived from proliferative epithelial cells of the ectoderm germ layer. During primary neurulation, the neuroectoderm (neural plate) elongates and bends at the medial hinge point at the ventral most aspect of the neural plate. The medial hinge point remains anchored to the notochord along the long axis of the developing embryo. The notochord provides important signaling molecules such as *sonic hedgehog* for the formation of the ventral spinal cord. The notochord eventually becomes the nucleus pulposus of the intervertebral discs. The lateral margins (neural folds) of the neural plate then elevate until they fuse dorsally, creating a central neural groove and eventually the neural tube[3,5,15–17] (**Fig. 1**). Under the influence of signaling