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Abstract

A 6-hour-old Quarter Horse colt was examined because of an inability to rise. An uneventful parturition after a normal gestational length from a multiparous mare was reported by the owner. The colt was administered dexamethasone (4 mg) and gentamicin (400 mg) IV and mare's colostrum (900 mL) PO before referral. Upon examination, the colt (59 kg) was depressed and recumbent. The rectal temperature was 97.6uF, heart rate was 110 beats/min, and the respiratory rate was 40 breaths/min. The colt had a prominent domed forehead, but no other significant abnormalities were detected on physical examination. Hematologic and biochemical abnormalities were limited to an increased hematocrit (53%; reference range, 37–49%), hypoproteinemia (4.3 g/dL; reference range, 5.1–7.6 g/dL), and an increased serum creatine kinase activity (1597 U/L; reference range, 65–380 U/L). Results of the arterial blood gas analysis were normal. Initial treatment included fluids (500 mL 0.45% NaCl IV q2h), mannitol (1 g/kg IV q12h), amikacin (25 mg/kg IV q24h), ceftiofur (10 mg/kg IV q12h), equine plasma (1000 mL IV once), and feeding via a nasoesophageal tube (500 mL mare's milk q2h).

Disciplines

Large or Food Animal and Equine Medicine | Veterinary Medicine | Veterinary Pathology and Pathobiology

Comments

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Dandy-Walker–Like Syndrome in a Quarter Horse Colt

David Wong, Matthew Winter, Joseph Haynes, Brett Sponseller, and Jennifer Schleining

A 6-hour-old Quarter Horse colt was examined because of an inability to rise. An uneventful parturition after a normal gestational length from a multiparous mare was reported by the owner. The colt was administered dexamethasone (4 mg) and gentamicin (400 mg) IV and mare's colostrum (900 mL) PO before referral. Upon examination, the colt (59 kg) was depressed and recumbent. The rectal temperature was 97.6°F, heart rate was 110 beats/min, and the respiratory rate was 40 breaths/min. The colt had a prominent domed forehead, but no other significant abnormalities were detected on physical examination. Hematologic and biochemical abnormalities were limited to an increased hematocrit (53%; reference range, 37–49%), hypoproteinemia (4.3 g/dL; reference range, 5.1–7.6 g/dL), and an increased serum creatine kinase activity (1597 U/L; reference range, 65–380 U/L). Results of the arterial blood gas analysis were normal. Initial treatment included fluids (500 mL 0.45% NaCl IV q2h), mannitol (1 g/kg IV q12h), amikacin (25 mg/kg IV q24h), ceftiofur (10 mg/kg IV q12h), equine plasma (1000 mL IV once), and feeding via a nasoesophageal tube (500 mL mare's milk q2h).

The colt became more alert and would struggle to rise intermittently over the next 12 hours and could briefly maintain a sternal position or stand when assisted. When standing, the colt would constantly sway his head and neck from side to side. Simultaneously, a pronounced truncal and hindquarter sway was observed in the same direction of head movements (See the video available online at <http://dx.doi.org/10.1892/06-290v1>). No abnormalities were noted for a foal of this age involving the cranial nerves, patellar reflex, triceps reflex, or withdraw/flexor reflex. There was neither opisthotonus or intention tremors. Precise assessment of proprioception and postural reactions was not possible, because the colt could not stand or ambulate on his own. Based on neurologic examination, a lesion involving the cerebellum was suspected.

On the second day of hospitalization, the colt was alert and would frequently struggle and make attempts to rise. There was a leukocytosis ($13.50 \times 10^3/\mu\text{L}$; reference range, $4.9\text{--}11.7 \times 10^3/\mu\text{L}$) characterized by a neutrophilia ($12.55 \times 10^3/\mu\text{L}$; reference range, $3.36\text{--}9.57 \times 10^3/\mu\text{L}$); these changes could have been induced

by stress, administration of steroids the day before or both. The serum immunoglobulin G concentration was greater than 800 mg/dL. A congenital malformation of the central nervous system (CNS) was suspected and computed tomography (CT) of the head was performed with the foal under general anesthesia. Transverse, contiguous 3-mm scans were obtained from the level of the frontal sinuses caudal to the first cervical vertebra. Studies were performed with and without administration of contrast (Hypaque^a 2 mL/kg IV). The 4th ventricle was markedly dilated, and, in the caudal fossa, dorsal to the rostral and caudal cerebellar peduncles in the region of the vermis, there was marked hypodensity (Figs 1 and 2). CT numbers in this location were consistent with (cerebrospinal fluid) (6–20), and markedly hypodense to the surrounding brain parenchyma (30–45). The lateral cerebellar hemispheres were considered normal, and there was no contrast enhancement. There were open sutures between the occipital and parietal bones, as well as the frontal and parietal bones, which were considered normal for a foal of this age. Findings were consistent with a hypoplastic vermis and ventriculomegaly of the 4th ventricle.

The colt was euthanized, and a postmortem examination was performed. Relevant findings were limited to the brain and the skull. Aplasia of the cerebellar vermis was confirmed; the lateral cerebellar hemispheres were present but were separated by a gap where the vermis typically is positioned (Figs 3 and 4A). In addition, the lateral ventricles were mildly dilated. The foramina that connect the lateral ventricles to the third ventricle were dilated, as was the mesencephalic aqueduct. The 4th ventricle was dilated and covered by a thin roof of fibrous tissue, which was fused with the meninges, forming a meningocele (Fig 4A). The cisterna magna was expanded with cerebrospinal fluid, the corpus callosum was absent (Fig 4B), and the leptomeninges of the cerebral hemispheres were somewhat fused together along the longitudinal fissure. Furthermore, the gyri of the cerebral hemispheres were small, more numerous than normal, and somewhat convoluted (polymicrogyria).

Microscopic findings of the cerebellar hemispheres consisted of dysplasia of the medial portions of the hemispheres that would normally be fused with the vermis; this area was characterized by few Purkinje neurons involving the molecular layer, whereas the granular layer was hypocellular and disorganized. The remainder of the cerebellar hemispheres was microscopically normal. Microscopic examination of the cerebral hemispheres, adjacent to the longitudinal fissure, revealed the absence of a corpus callosum, with small gyri that were more numerous than normal (polymicrogyria); the cerebral cortex was somewhat thinner in these areas. There were no gross or microscopic abnormalities of the spinal cord.

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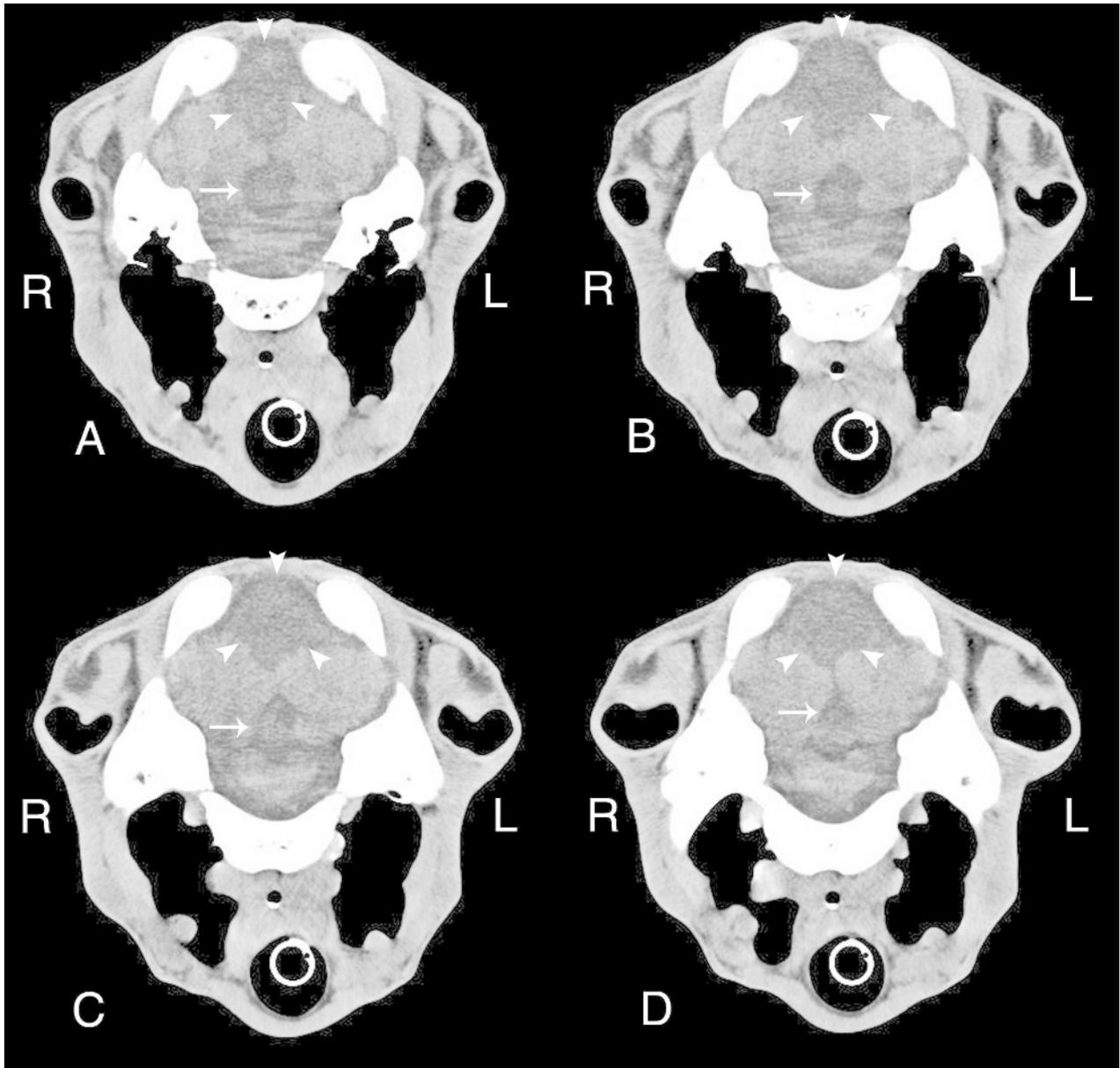


Fig 1. (A–D) Transverse, contiguous 3-mm scans of the caudal fossa displayed in a brain window depicting the absence of the cerebellar vermis (arrowheads) and the enlargement of the 4th ventricle (arrows). Also note the open suture at the level of the occipital bones. This is considered normal for a foal of this age.

The reported incidence of congenital brain malformations in horses is relatively low. Hydrocephalus is the most commonly reported congenital malformation of the brain, with an incidence of 3% in 1 study involving 608 fetuses or foals presented for postmortem examination.¹ Less common disorders reported in fetuses or foals include cerebellar abiotrophy; cerebellar aplasia, or hypoplasia; hydrancephaly; anencephaly; and merocrania.^{2–4} The clinical signs, diagnostic imaging, and postmortem examination findings in this case represent a constellation of abnormalities known as Dandy-Walker-like syndrome (DWS) in people. DWS has been reported in calves, lambs, dogs, goats, a kitten, and a foal.^{5–10} The current foal had clinical signs and postmortem findings different than the previously

reported case, likely representing the heterogeneous group of lesions and clinical signs that can be associated with DWS, as described for people.⁵ Core abnormalities of DWS in people encompass 3 pathologic criteria: (1) cerebellar vermian hypoplasia or agenesis, (2) 4th ventricle dilation, and (3) communicating hydrocephalus with enlarged lateral ventricles.^{8,11} Agenesis or dysgenesis of the corpus callosum is the most commonly associated CNS malformation in people with DWS.⁸ Systemic manifestations associated with DWS in people include malformations of the cardiovascular (ventricular septal defects), urogenital (hydrocele), intestinal (duodenal atresia), facial (cleft palate), and musculoskeletal (malformed limbs) systems.¹²

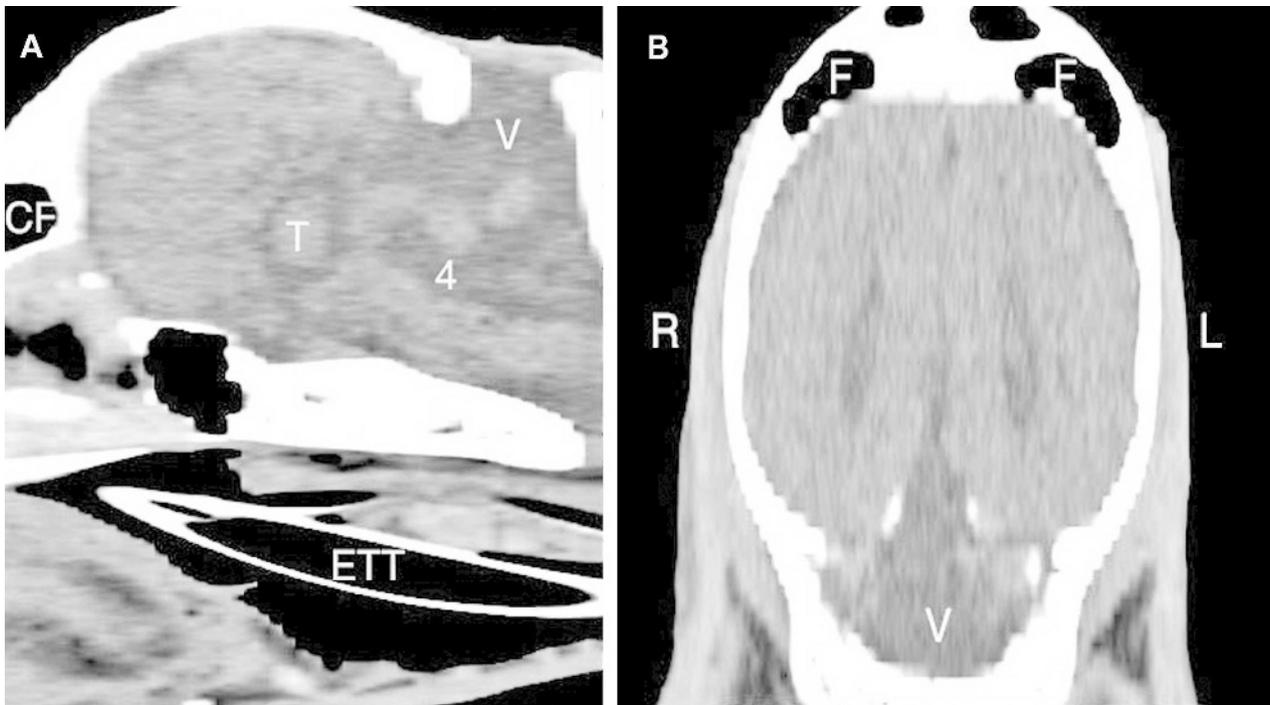


Fig 2. (A) Sagittal reformatted image of the cranial vault displayed in a brain window and depicting the absence of the cerebellar vermis (V) and the enlargement of the 4th ventricle (4). Also note the open suture at the level of the occipital and parietal bones. Rostral is to the left of the image. Conchofrontal sinus (CF); interthalamic adhesion (T); 4th ventricle (4); region of the cerebellar vermis (V); endotracheal tube (ETT). (B) Dorsal plane reformatted image of the cranial vault displayed in a brain window displaying the absence of the cerebellar vermis (V). Rostral is at the top of the image. Frontal sinus (F); region of cerebellar vermis (V).

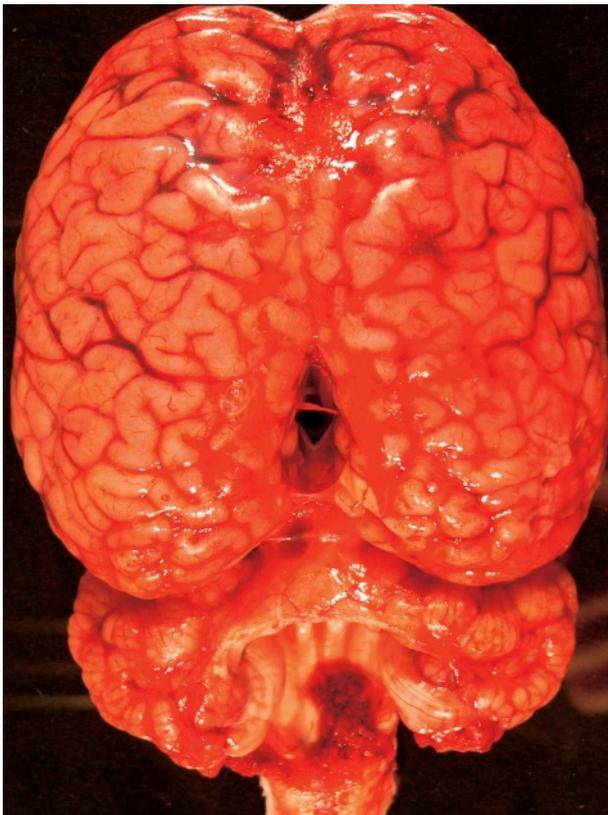


Fig 3. Entire brain showing apparent fusion of the medial aspects of the cerebral hemispheres and lack of the cerebellar vermis.

The cerebellum regulates and coordinates fine motor responses and maintains equilibrium and normal body position at rest and during movement.¹³ Because the degree of cerebellar hypoplasia can range from mild vermian hypoplasia to nearly complete absence of the cerebellum, clinical signs are variable in people with DWS.¹⁴ In the foal reported here, the head and truncal swaying and the inability to stand up because of poor balance control were suggestive of cerebellar disease. The cerebellum can be organized into longitudinal zones, which include the vermian, intermediate, and lateral zones. Considerable overlap in the functions of each zone exists, but, in general, abnormalities of the vermian zone, as noted in the foal here, result in disorders of stance, gait, movements of the eyes and the body with respect to gravity and movement of the head in space.¹⁵ Therefore, the clinical signs noted in the foal were attributed to vermian aplasia associated with DWS. CT was a valuable antemortem diagnostic tool that facilitated the diagnosis of DWS. CT clearly demonstrated the absence of a normal vermis and the dilation of the 4th ventricle; these findings were confirmed on postmortem examination. However, the absence of the corpus collosum and the mild dilatation of the lateral ventricles noted at postmortem examination was not identified on CT.

Similarities between the previously described foal with DWS and the current foal include the inability to maintain sternal recumbency and the inability to stand

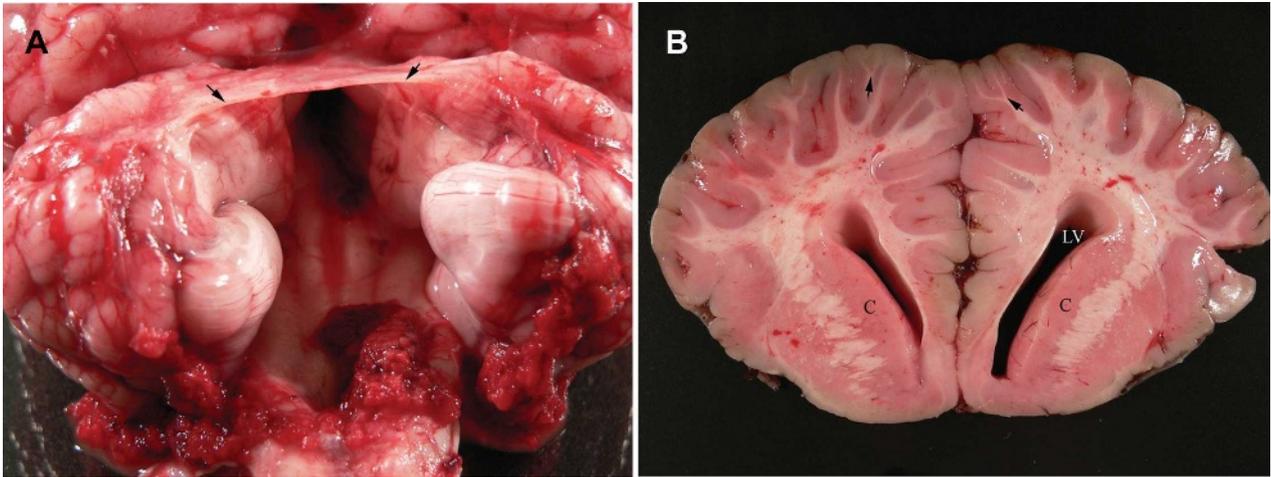


Fig 4. (A) The 4th ventricle is evident because of a complete lack of the cerebellar vermis. There is a fibrous membrane covering the roof of the 4th ventricle (arrowheads); this was fused with the meninges forming a meningocele. (B) Cross-section of the brain at the level of the caudate nuclei (C) revealing a lack of the corpus callosum, mild dilatation of the lateral ventricle (LV), and polymicrogyria (arrowheads).

voluntarily. However, differences in clinical signs outnumbered similarities. The foal reported here demonstrated head, truncal, and hindquarter swaying but was relatively bright and responsive, whereas the previously reported foal did not have truncal sway and was lethargic, occasionally demonstrating periods of unconsciousness. The previously reported foal had additional deficits, including nystagmus, intention tremor, wide-based stance and gait, aggressive behavior, and abnormal respiratory pattern. Abnormal respiratory patterns have been associated with DWS; however, seizures are an uncommon clinical sign.⁵ The investigators of that report suggested that the foal's seizure activity and behavioral changes were indicative of a concurrent but undefined, forebrain disorder. However, changes in mentation have been attributed to increased intracranial pressure in people and a kitten with DWS, and may have been a contributing factor.^{8,10}

Anatomic differences between this case and the previously reported foal include polymicrogyria in contrast to good gyral pattern and complete aplasia of the cerebellar vermis in contrast to aplasia of the caudal two thirds of the cerebellar vermis, respectively.⁵ The frontal poles, caudate nuclei, and corpora striatum appeared bulbous in the previously reported foal, along with the absence of the rostral commissure, caudal commissure, and nodulus, and the presence of irregular cerebellar folia. Similarities between the 2 foals include the absence of the corpus callosum, the presence of both cerebellar hemispheres separated by a gap where the vermis is normally located and dilatation of the 4th ventricle.

Clinical signs in other species are similar to many of those noted in the foal. Puppies with DWS demonstrate ataxia, intention tremors, nystagmus, circling, wide lateral head excursions, and truncal sway, whereas calves and lambs may be born dead or may demonstrate difficulty standing and severe incoordination.^{6,7,9,16} A kitten with DWS demonstrated lethargy, an inability to

stand or walk, nystagmus, and postural deficits, whereas people may have cognitive delay, apnea, seizures, ataxia, hypotonia, spasticity, nystagmus, and opisthotonus.^{8,10,11,14} Although clinical signs are variable among individuals with DWS, an apparent similarity is the presence of a domed cranium (macrocephaly).^{5,6,9,11,14}

The cause of DWS is unknown, although multiple theories involving infectious, toxic, and genetic variables have been proposed. Embryonic cerebellar development arises from 2 distinct primordia known as rhombic lips. In human fetuses, proliferating neuroblasts from symmetric alar plates form paired rhombic lips that thicken, project into the 4th ventricle, and extend progressively toward midline.¹⁷ The rhombic lips on the 2 sides begin to fuse at the midline, starting rostrally, to form the cerebellar vermis primordium.¹⁸ Based on embryologic formation of the cerebellum, DWS is believed to result from an insult to the alar plate involving the dorsal 4th ventricle and rhombic lips. The precise gestational time in which embryonic formation of the cerebellum of the developing equine fetus is not known but appears to develop in a well-ordered sequence of events, similar to the human fetus.¹⁸ Currently, the authors of this report have no clear etiology as it relates to DWS in horses but speculate that an extensive and distinct insult at a specific time period of equine fetal development of the cerebellum results in the anatomic abnormalities. Other possible causes include in utero viral infections, which have been associated with cerebellar malformation in multiple species. These include bovine viral diarrhea virus in calves,¹³ panleukopenia virus in kittens,¹⁹ Cache Valley virus in lambs,²⁰ and hog cholera virus in piglets;¹³ virally induced cases have not been reported in foals.⁵ Histopathologic lesions associated with DWS do not resemble a viral or bacterial agent, nor has any infectious agent been identified from fetal CSF, amniotic fluid, or brain tissue in any species with DWS, making an infectious cause unlikely.^{6,16,21} A genetic component

resulting in DWS is possible but the occurrence of DWS among human siblings is exceptionally low.¹⁴ No clear toxic agent has been identified in any species.⁹

Footnotes

^a Hypaque, diatrizoate sodium injection, Amersham Health, Princeton, NJ

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