CASE PRESENTATION

Congenital Hydrocephalus in a Neonatal Foal*

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A 3-day-old, full-term Thoroughbred colt (weight: 121 lb [55 kg]) was admitted to the Veterinary Medical Teaching Hospital, University of Wisconsin–Madison, for evaluation and treatment of hypoxic ischemic encephalopathy. The colt was born without assistance and without complications at delivery. It was initially unable to nurse from the dam and was bottle-fed during the first 3 days of life. On the first day of life, the colt chewed constantly and developed a head tilt and a propensity to hold its nose straight up in the air or suck on its own legs. For the first 2 days of life, the colt was initially treated with balanced polyionic fluids, hyperimmune equine plasma, gentamicin, penicillin, flunixin meglumine, omeprazole, and dimethyl sulfoxide, which were all given intravenously at recommended doses.

At presentation at the University of Wisconsin–Madison, the colt was ambulatory but lethargic with no suckle reflex. The colt appeared to be unaware of its surroundings and oblivious to the mare. Its respiratory rate was 39 breaths/min, heart rate was 80 bpm, and rectal temperature was 101.5˚F (38.6˚C). Ophthalmic examination revealed a large, superficial corneal erosion in the right eye and bilateral and symmetric posterior cortical cataracts. During cardiac auscultation, the foal had a left basilar systolic murmur consistent with patent ductus arteriosus. A small umbilical hernia was noted, as was tendon flexor laxity in all four limbs. No other physical examination abnormalities were detected.

Results of a complete blood cell count were within reference ranges. Abnormalities in the serum biochemical profile included hypoalbuminemia (2.4 g/dl; reference range: 2.8 to 3.7 g/dl)¹ and increased creatinine kinase (444 U/L; reference range: 21 to 97 U/L).¹ Serum IgG concentrations as measured by radial immunodiffusion were 861 mg/dl (normal range: >800 mg/dl). The sepsis score was zero.

TREATMENT AND CLINICAL COURSE

Initial therapy consisted of intravenous administration of potassium penicillin (22,000 IU/kg q6h), amikacin (20 mg/kg q24h), and thiamine (10 mg/kg q12h). Two liters of plasma containing 2,000 U/L of heparin were administered at 100 ml/hr for 30 minutes and then at 250 ml/hr. A 10% magnesium sulfate solution (33,000 mg) was administered at 45 ml/hr IV. A 10% dextrose solution (5 g/hr dextrose) was also administered intravenously for 4 days. The antibiotics and thiamine were given during the 5 days of treatment. Omeprazole (4 mg/kg PO q24h) was given, and a triple antibiotic ophthalmic ointment was applied to the right eye every 8 hours.

The colt nursed from a bottle every 2 hours, receiving 14% of its body weight per day, and was encouraged to nurse from the dam. During the following 5 days, the colt’s

*A case commentary begins on page 108.
neurologic status did not significantly improve. The colt wandered aimlessly, frequently bumping into inanimate objects, and showed no normal bonding activity with the dam. Because of the clinical signs and poor response to treatment, plain radiography of the skull and computed tomography (CT) were conducted to further evaluate skull and intracranial anatomy. Findings on lateral and dorsoventral skull radiographs were within normal limits. CT showed asymmetric (i.e., left larger than right), severe hydrocephalus with slight deviation of the midline to the right side. The mesencephalic aqueduct and third ventricle appeared dilated, and accumulation of increased amounts of cerebrospinal fluid (CSF) surrounded the brain and brain stem (Figure 1).

Figure 1. CT scan of the brain. Comparison between a foal with hydrocephalus (A and B) and a normal age-matched foal (C and D). Note the deviation of the midline to the right side and marked dilation of the lateral ventricles (arrow in A) compared with the normal ventricular dimensions shown in C (arrow). Dilation of the ventricular system and mesencephalic aqueduct is also evident at the level of the third ventricle in the hydrocephalic foal (arrows in B).
Because of the grave prognosis for the colt, the owners elected euthanasia. Necropsy revealed that the calvarium was asymmetric (i.e., left skullcap: $5 \times 10$ cm; right skullcap: $4.3 \times 10$ cm). There were marked abnormalities of the gyri and sulci of the cerebral cortex, with dimpling cranially and decreased sulci and coalescing gyri caudally (Figure 2). The lateral ventricles were markedly dilated, resulting in severe cerebrocortical thinning (average thickness: 1.5 cm). The third ventricle and mesencephalic aqueduct were markedly dilated (i.e., $2 \times 1$ cm and 0.8 cm in diameter, respectively). The fourth ventricle was moderately dilated (Figure 3).

DISCUSSION

The terms hypoxic ischemic encephalopathy, peripartum asphyxia syndrome, and neonatal maladjustment syndrome are used to describe noninfectious central nervous system (CNS) disorders related to hypoxia in newborn foals before, after, or during delivery, respectively. Several neurologic conditions with different causes have been included in this category, so more specific terminology has to be used to describe neurologic diseases in foals. In addition to hypoxic ischemic encephalopathy, other common causes of neurologic signs and seizures in newborn foals are CNS congenital abnormalities, trauma, bacterial meningitis, septicemia, and idiopathic epilepsy. Hypoxic ischemic encephalopathy and congenital abnormalities are commonly accompanied by septicemia because lack of nursing during the first 24 hours of life predisposes foals to failure of passive transfer and infection.\(^5\)

In this foal, there was no corroborating evidence of infection or failure of passive transfer because the complete blood cell count and serum IgG concentrations were within normal limits and the sepsis score was zero. The colt received supportive treatment, plasma, and antibiotics as prophylaxis for a compromised neonate. Bilateral posterior cataracts were symmetric and likely congenital.

In general, hydrocephalus may be classified as being either compensatory or obstructive. Compensatory (normotensive) hydrocephalus commonly develops as a result of a viral infection or another infectious agent in utero and is also called hydranencephaly. Destruction of brain tissue results in severe accumulation of CSF.\(^6\) In this form of hydrocephalus, the CSF pressure is usually not increased and CSF accumulates in areas where brain tissue has been injured or inflamed. In contrast, the obstructive (hypertensive) form of hydrocephalus occurs as a result of obstruction to CSF outflow or impairment in CSF absorption. This condition has been previously recognized in Arabian foals.\(^6-8\) Animals with hydrocephalus often are born dead or die shortly after birth. The most common clinical signs are depression, failure

Figure 2. Macroscopic view of the brain and calvarium. Decreased sulci and coalescing gyri in the caudal portion of the cerebral cortex (arrow).

Figure 3. Cross-section through the rostral cerebral hemispheres showing marked dilation of the lateral ventricles (A and B, arrows) and significant dilation of both caudal lateral ventricles, the mesencephalic aqueduct, and the third ventricle (C and D, arrows). Note the severe cortical thinning due to dilation and accumulation of CSF in the ventricles.
Making the Rounds

108

Making the Rounds

to bond with the dam, partial failure of suckling, conscious proprioceptive deficits, blindness, and head tremors. Affected foals often do not nurse sufficient amounts of colostrum and die from septicemia.

The case reported here is notable because of the advanced diagnostic imaging procedure performed to evaluate the calvarium, brain, and proximal vertebral cord in a foal with altered mentation that was refractory to conventional treatment for hypoxic ischemic encephalopathy. Although conventional radiography can be helpful in diagnosing skeletal trauma involving the basilar region of the equine skull, superimposition of bony structures and difficulties in evaluating small differences in tissue density limit the usefulness of survey radiography. Furthermore, the location and extent of both intracranial and calvarial lesions are difficult to define by radiographic examination. Sensitive differentiation of tissue physical densities is best obtained by CT, which permits detection and localization of intracranial structural lesions, such as hydrocephalus, intracranial hemorrhage, and brain trauma.

Magnetic resonance imaging provides superior contrast resolution of soft tissue and is the premiere imaging modality in diagnosing intracranial CNS lesions. In this case, CT was considered the first diagnostic option because of its reduced cost compared with magnetic resonance imaging. Severe hydrocephalus, which is probably congenital in origin, was diagnosed by CT and confirmed by necropsy. This case report highlights the value of considering CT in identifying structural, congenital, and intracranial soft tissue lesions in newborn foals. We suggest that this diagnostic modality be considered in foals with neurologic signs consistent with an intracranial lesion, particularly those that are refractory to conventional therapy for hypoxic ischemic encephalopathy or sepsis.

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References


Case Notes and Commentary

Congenital Hydrocephalus

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Hydrocephalus is uncommon in horses and is usually identified as a neonatal problem and/or diagnosed at necropsy. A review of case records for New Bolton Cen-
of PubMed. Most reported cases were described as congenital defects. The reported prevalence of congenital and infantile hydrocephalus in humans is 0.48 to 0.81 per 1,000 births (live and still), and a large number of those patients have significant neurologic and developmental deficits. Although congenital hydrocephalus is primarily considered a sporadic event, familial and heritable forms of the disease have been suggested in horses.

A generally accepted view of cerebrospinal fluid (CSF) circulation is that CSF is produced by the choroid plexus and flows to the pacchionian granulations, where reabsorption occurs as a passive process. According to this view, CSF flow is driven by a pressure gradient between the subarachnoid space and the major dural venous sinuses within the cranial cavity. A more recent alternative theory is that CSF absorption primarily occurs through the blood capillaries. In “communicating” hydrocephalus (i.e., restricted arterial pulsation hydrocephalus), CSF flow across the foramen magnum is significantly impeded by arterial pulsation during systole. Conversely, in “obstructive” hydrocephalus (i.e., venous congestion hydrocephalus), it is argued that ventricular dilation compresses the cortical veins, resulting in increases in cerebral blood volume and intracranial pressure. Most diseases resulting in hydrocephalus occur by interfering with CSF reabsorption. Although discussion of CSF dynamics may seem to be of only academic interest, distinguishing between “communicating” and “obstructive” hydrocephalus is clinically important in infants because treatment options differ, depending on the underlying cause of the condition.

Causes of infantile and congenital hydrocephalus described in horses are not numerous. The most easily diagnosed are those associated with meningomyelocele as reported in miniature horses in association with spina bifida, in which concurrent soft tissue swellings in the head and neck region and neurologic abnormalities should increase suggestion of hydrocephalus. Clinical choices for management are not difficult in such cases, with euthanasia frequently being chosen once the diagnosis is clearly confirmed. Cases similar to that presented in the accompanying case report, in which advanced diagnostic modalities are required, are more diagnostically challenging. Because normal computed tomography (CT) reference images have been produced for foals, the utility of this technique in aiding diagnosis of neurologic abnormalities has been improved. Diagnosis of unusual presentations of neurologic abnormalities in neonatal foals are frequently ascribed to hypoxic ischemic encephalopathy, which is also known as neonatal encephalopathy. Consequently, considerable time and financial resources can be spent futilely pursuing intensive care and supportive therapies.

Once the decision has been made to refer an equine neonate with neurologic abnormalities to an equine neonatal intensive care unit or other facility capable of providing the appropriate level of care, owners have generally opted for significant financial investment. In cases in which the presentation is unusual and the response to therapy is atypical, expensive diagnostic modalities (e.g., CT or magnetic resonance imaging) may, in fact, be quite reasonable. Use of such techniques may provide information that allows owners to make decisions regarding continuing expensive treatment in a timely manner. Facilities offering these diagnostic imaging modalities are increasing in number, and the availability of mobile units in certain regions of the country allows remote clinics to offer these diagnostic aids. CT (continues on p. 110)
Making the Rounds (continued from p. 109)

and magnetic resonance imaging should be considered for evaluating foals such as the one described in the accompanying case report.

REFERENCES