# Congenital porencephaly with cerebellar hypoplasia in a Holstein calf: a case report

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**ABSTRACT**: We describe the case of a nine-day-old female Holstein calf which had cheiloschisis, a moderate dome-shaped head, ataxia and opisthotonus since birth. No significant findings except the dome-shaped head were observed on survey radiography of the skull. Computed tomography (CT) images showed bilateral lateral ventriculomegaly, cerebellar hypoplasia and a cyst-like lesion communicating with the right lateral ventricle. Post-mortem examination revealed a cerebral defect in the frontoparietal lobe, which communicated with the right lateral ventricle, and cerebellar hypoplasia. CT provided a characteristic finding of porencephaly and was helpful for diagnosing the accompanying anomalies. We suggest that porencephaly should be included as a specific anomaly in the differential diagnosis of congenital brain malformation.

**Keywords**: accompanying anomalies; congenital malformation; computed tomography; diagnosis; calf

Porencephaly is an uncommon neurological disease characterized by the presence of a cerebrospinal fluid-filled cavity or cavities within the brain (Gould et al., 2005). Congenital anomaly (Lee et al., 2009), intrauterine infection (Kirkland et al., 1988; Hewicker-Trautwein et al., 1995) and prenatal ischemia (Denis et al., 2000) are etiological factors for porencephaly in domestic animals. Therefore, porencephaly manifests in young animals. In addition, congenital porencephaly may be accompanied by other anomalies such as hydrocephalus, arachnoid cyst, optic chiasm agenesis and optic nerve hypoplasia (Lee et al., 2009). Hydrocephalus, one of the accompanying anomalies, has a positive correlation with short stature or slow growth in humans (Klauschie and Rose, 1996). A previous study reported that sheep with porencephaly due to bluetongue virus infection showed retarded growth (Richardson et al., 1985). On the basis of these studies, it was postulated that cattle with porencephaly, with or without accompanying anomalies, may display reduced growth, which could eventually result in economic losses.

Computed tomography (CT) and magnetic resonance (MR) imaging are widely used for the diagnosis of porencephaly in human medicine (Raybaud, 1983; Shen et al., 1993). However, porencephaly is usually diagnosed by necropsy and histopathological examination in veterinary practice (Scholes et al., 2009). There are only two reports in the veterinary literature of porencephaly diagnosed by imaging in a dog (Utsunomiya, 1982) and a calf (Lee et al., 2009). This report describes the clinical signs and CT characteristics of porencephaly and accompanying anomalies in a calf.

## Case description

A nine-day-old female Holstein calf was referred with cheiloschisis and ataxia since birth. Physical examination revealed a moderate dome-shaped head, cheiloschisis, ataxia and intermittent opisthotonus. Neurological examination revealed right-sided head turn and decerebellate rigidity. The menace response was decreased in both eyes,

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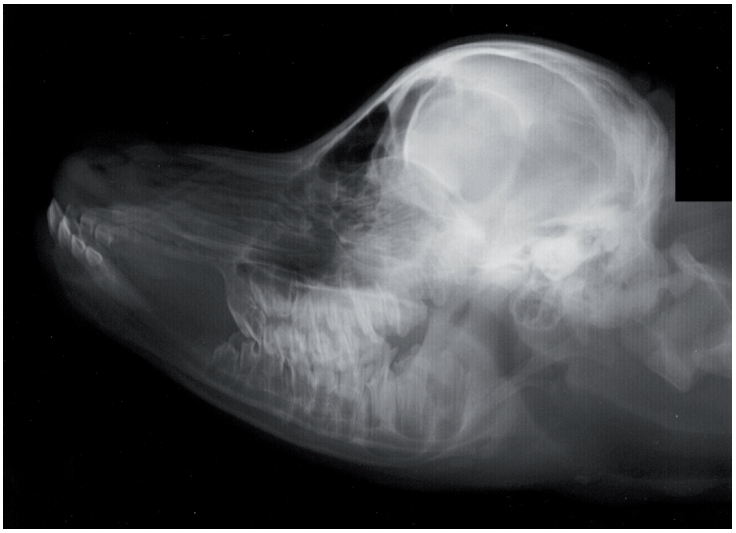

Figure 1. Lateral radiography of the skull. No significant observations except the dome-shaped cranium were made

but vision and other cranial nerves were appropriate. Spinal reflexes and pain perception were normal. Based on the neurological examination findings, the lesion was localized to the prosencephalon and cerebellum. Differential diagnoses included hydrocephalus, a complex brain anomaly such as Dandy-Walker malformation, meningoencephalitis and neoplasia.

Complete blood cell count and the serum biochemistry profile were within the reference range. Protein and cell counts in the cerebrospinal fluid, collected by lumbar puncture, were normal. No significant findings except the dome-shaped head were observed on survey radiography of the skull (Figure 1). CT of the head was performed using four detector-row CT (Asteion Super 4, Toshiba, Japan) under general anaesthesia. The calf was positioned in ventral recumbency on the CT table. Contiguous CT images were obtained with a slice thickness of 2.0 mm, 120 kV, 150 mA and tube rotation time of 0.75 s. The CT images were reconstructed on an image-processing workstation (Virtual Place, AZE, Japan) and evaluated with three cross-sectional images (transverse, sagittal and dorsal planes). Bilateral lateral ventriculomegaly and a cyst-like lesion within the right cerebrum (5.0 Hounsfield units) were evident on non-contrast CT images; the cavity was situated in the right frontopari-

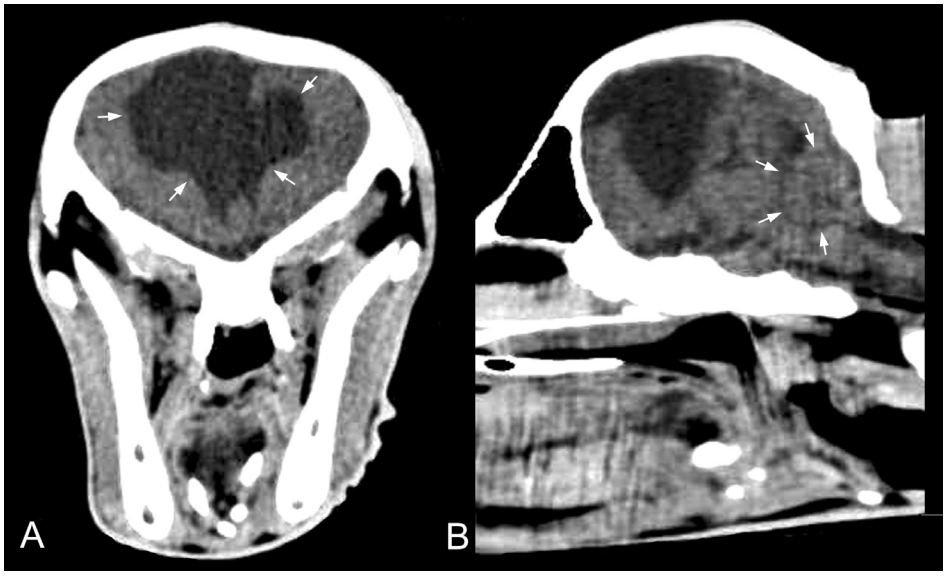


Figure 2. Non-contrast transverse (A) and sagittal (B) computed tomographic images of the brain. Bilateral ventriculomegaly and a cyst-like lesion (A, arrows) communicating with the right lateral ventricle are observed. Note the absence of the septum pellucidum. The cerebellum is decreased in size (B, arrows)

etal region, communicated with the right lateral ventricle (Figure 2A) and extended caudally to the cerebellum. The cerebellum was decreased in size (Figure 2B). Septum pellucidum was absent. In addition, a small rostrocerebellar diverticulum and a cleft in the middle of the upper lip were observed.

The calf was euthanized after the CT examination because of poor prognosis. There was bilateral dilation of the lateral ventricles. A cerebral defect was evident in the right frontoparietal region, communicating with the right lateral ventricle. The wall of the cleft consisted of white matter with no cortical gray matter. There was clear hypoplasia of the entire cerebellum (2.5 × 3.0 cm) (Figure 3). The additional gross post-mortem finding was a cleft in the middle of the upper lip. Rostrocerebellar diverticulum observed in CT images was not detected on post-mortem examination. This may due to the size of the rostrocerebellar diverticulum. The lesion was small and deflated after the brain was removed. Histopathologically, no abnormalities were observed in the central nervous system except cerebellar hypoplasia. No abnormalities were observed in any other organs. A diagnosis of porencephaly with cerebellar hypoplasia was made based on CT and histopathological findings.

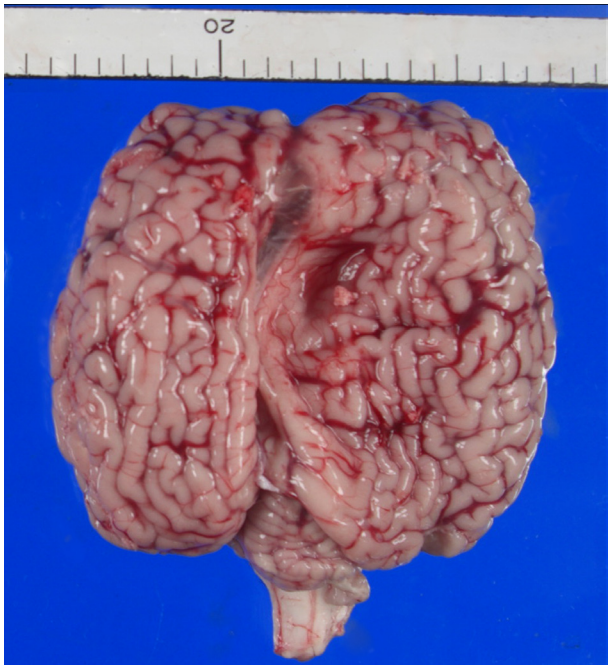


Figure 3. Photograph of the brain. The cerebral defect can be seen in the frontoparietal region. Note the obvious hypoplasia of the entire cerebellum

## DISCUSSION AND CONCLUSIONS

Animals with congenital porencephaly present with neurological signs at birth and these neurological signs depend on the location of the porencephalic defects (Lee et al., 2009). The case presented here had non-progressive neurological signs, including ataxia and head turn at birth. Furthermore, the head was dome-shaped, which has been reported in association with hydrocephalus in animals (Bagley, 2004). This report suggests that porencephaly should be considered in the differential diagnosis of neurological signs at birth with or without a dome-shaped head.

Porencephaly is characterized by a cavity or cleft within the brain that is not lined by gray matter (Barkovich and Norman, 1989). Computed tomographic imaging demonstrates the porencephalic cavity within the cerebrum (Barkovich and Norman, 1988; Byrd et al., 1989), however, MR imaging provides better delineation of anatomic detail and may detect more subtle comorbid brain anomalies. In the present case, the CT imaging obtained using an Asteion Super 4 model scanner failed to delineate the lining of the cavity. This did not preclude diagnosis of porencephaly because of the location of the cavity within the cerebrum in clear communication with the lateral ventricle.

In humans, unifocal porencephalic defects are mainly located in the frontoparietal region, whereas multiple defects are present in the basal ganglia (Kolawole et al., 1987). The extent of porencephaly does not correlate with the severity of clinical signs in people (Ho et al., 1998). In a previous report of porencephaly in a calf, a unifocal defect was located in the frontoparietal region (Lee et al., 2009) and in the present case the defect was located in the same area suggesting an anatomic predilection for unifocal porencephaly in this region of the brain. This hypothesis is supported by the observation that solitary porencephalic lesions are mainly located in the frontoparietal region in humans (Kolawole et al., 1987); however, more studies are needed in animals to confirm a predisposition for porencephaly in this area.

Differential diagnoses of porencephaly include schizencephaly and hydranencephaly. Schizencephaly is characterized by a gray matter-lined cleft that extends from the pial surface of the cortex to the lateral ventricle(s) (Denis et al., 2000). Schizencephaly is typically a bilateral defect (Raybaud, 1983), although unilateral defects have

also been reported (Denis et al., 2000). Unilateral schizencephaly may be confused with porencephaly in CT or MR imaging if the cleft cannot be clearly seen to originate from the surface of the brain or the lining of the cleft cannot be determined (Denis et al., 2000). Hydranencephaly develops as a result of the same encephaloclastic processes that produce porencephaly; however, the lesion is much more severe. In cases of hydranencephaly there is complete loss of brain parenchyma leading to a large fluid filled cavity within the brain that is covered by a pial-glial membrane (Barkovich and Norman, 1989; Stevenson et al., 2001). Hydranencephaly is classified as a type of porencephaly (Barkovich and Norman, 1989).

Congenital porencephaly is often accompanied by other anomalies (Stewart et al., 1978; Ho et al., 1998; Moinuddin et al., 2003). Hydrocephalus (Ho et al, 1998; Lee et al., 2009), intracranial arachnoid cysts (Lee et al., 2009), vascular anomalies (Stewart et al., 1978), intracranial haemorrhage (Moinuddin et al., 2003), optic chiasm agenesis (Lee et al., 2009), optic nerve hypoplasia (Lee et al., 2009) and corpus callosum agenesis (Lee et al., 2009) have been seen in association with porencephaly in humans and animals. In the calf presented here, a porencephalic defect was identified along with hydrocephalus and cerebellar hypoplasia. In addition, the calf had cheiloschisis and a dome-shaped head that was detected on physical examination. This report suggests that congenital porencephaly may be accompanied by other anomalies, similar to a previous study in animals, and that congenital porencephaly should be considered when congenital anomalies are detected at physical examination.

In the present study, CT was useful for the antemortem diagnosis of porencephaly in a calf. Based on a previous study (Lee et al., 2009) and the present case, congenital porencephaly in calves may be accompanied by other anomalies. We suggest that porencephaly be included as a specific anomaly in the differential diagnosis of congenital brain malformations in calves.

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