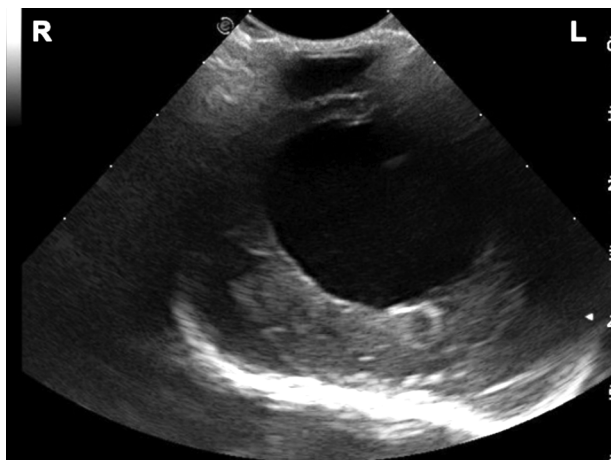


## What Is Your Diagnosis?

In collaboration with the American College of Veterinary Radiology



**Figure 1**—Transverse plane ultrasonographic image of the brain performed through the bregmatic fontanelle in a 4-month-old sexually intact Yorkshire Terrier examined because of abnormal behavior and visual impairment since adoption. The dorsal aspect of the head is toward the top of the image. L = Left. R = Right.

### History

A 4-month-old sexually intact male Yorkshire Terrier was examined because of a history of abnormal behavior and visual impairment since the dog was adopted from an animal shelter. On physical examination, the dog showed a large dome-shaped calvarium with 2 open fontanelles on the frontoparietal suture (bregmatic fontanelle) and ventrolateral bilateral strabismus. Abnormal findings on neurologic examination included altered mental status ranging from depression to hyperexcitability, reduced postural reactions in all four limbs, and blindness with absent menace response in both eyes. Neurologic signs were consistent with a lesion in the prosencephalic region. Results of hematology, biochemistry, and urinalysis

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were within the reference values. An ultrasonographic study of the brain was performed through a persistent dorsal midline fontanelle (**Figure 1**).

Formulate differential diagnoses, then continue reading.

### Diagnostic Imaging Findings and Interpretation

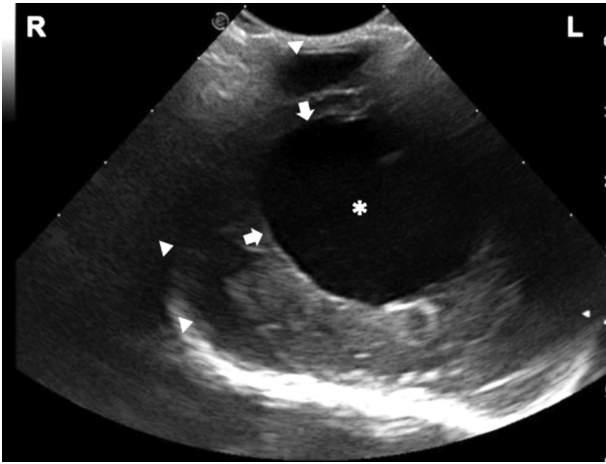
On ultrasonography, the lateral ventricles appeared enlarged and fused as a single cavity, and the brain parenchyma was surrounded by a large amount of an anechoic fluid (**Figure 2**). Pre- and postcontrast CT of the head was performed with a 16-slice helical multidetector CT scanner with a 1.25-mm slice thickness, 120 kVp, and 200 mA (Hitachi Eclis 16) (**Figure 3**). There was a moderate accumulation of fluid, isoattenuating to cerebrospinal fluid (CSF), between the skull and the brain cortex, considered to be an enlarged subarachnoid space. The cerebral brain parenchyma was smooth, narrow, and ring shaped, but no integrity defects were observed. The septum pellucidum was absent, and the lateral ventricles appeared enlarged and fused as a single cavity. Widened and open fontanelles were evident. Two small CSF samples were obtained, one obtained from the cerebellomedullary cistern and the other through the bregmatic fontanelle. Both CSF samples showed a nucleated cell count of < 5 cells/ $\mu$ L, and the protein concentration was within the reference limit. No organisms were apparent on cytopathological examination, but there was not enough sample to perform a bacterial culture. Based on these findings, a presumptive diagnosis of congenital internal and external hydrocephalus was made.

### Treatment and Outcome

The owner declined surgical placement of a ventriculoperitoneal shunt, and medical treatment with prednisone (0.5 mg/kg, PO, q 24 h) was started. Six months after diagnosis, on telephone interview, the owner described a mild improvement of clinical signs with a reduction of episodes of hyperexcitability but with persistence of visual impairment.

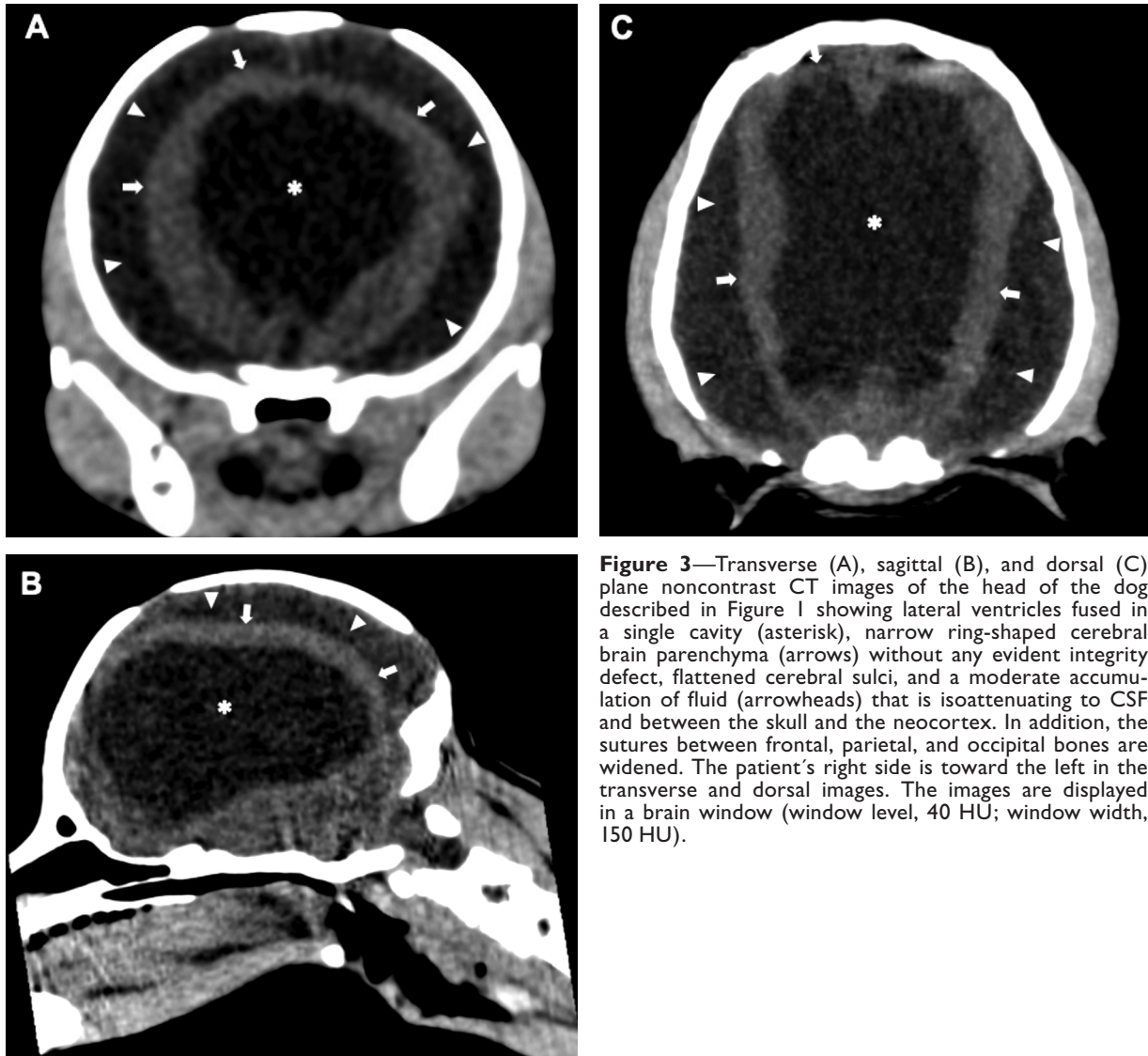
### Comments

Hydrocephalus is defined as an active distension of the ventricular system of the brain that results from



**Figure 2**—Same image as in Figure 1. The lateral ventricles appear enlarged and fused as a single cavity (asterisk), the brain parenchyma (arrows) was narrow and ring shaped, and the brain was surrounded by a large volume of anechoic fluid (arrowheads).

inadequate movement of CSF from the point of production within the ventricles to its point of absorption.<sup>1</sup> There are numerous classification schemes for hydrocephalus, and depending on the location of the accumulated CSF, hydrocephalus is classified as internal or external.<sup>1</sup> Internal hydrocephalus refers to increased fluid accumulation within the ventricular system and is one of the most common congenital anomalies of the nervous system in dogs.<sup>1</sup> In human medicine, the term external hydrocephalus is used to describe enlargement of the subarachnoid space with or without mild to moderate ventricular dilatation, although in a recent study, most patients had dilated lateral ventricles in addition to excessive external fluid on neuroimaging.<sup>1,2</sup> External hydrocephalus is occasionally described in humans,<sup>2</sup> but it was only reported in 3 dogs<sup>3-5</sup> and 2 cats.<sup>6</sup> Although the pathogenesis of external hydrocephalus is unknown, the most common theories suggest that external hydrocephalus is caused by either a congenital or acquired



**Figure 3**—Transverse (A), sagittal (B), and dorsal (C) plane noncontrast CT images of the head of the dog described in Figure 1 showing lateral ventricles fused in a single cavity (asterisk), narrow ring-shaped cerebral brain parenchyma (arrows) without any evident integrity defect, flattened cerebral sulci, and a moderate accumulation of fluid (arrowheads) that is isoattenuating to CSF and between the skull and the neocortex. In addition, the sutures between frontal, parietal, and occipital bones are widened. The patient's right side is toward the left in the transverse and dorsal images. The images are displayed in a brain window (window level, 40 HU; window width, 150 HU).

deficiency of the arachnoid villi in their ability to absorb the CSF that is produced continuously.<sup>2</sup> This results in distension of the ventricles and subarachnoid space. An alternate theory suggests that in patients with a severe internal hydrocephalus CSF accumulation within the lateral ventricle could lead to rupture of the cerebral parenchyma.<sup>2</sup> Our patient had severe dilatation of all the ventricles and the subarachnoid space, without inflammatory changes in the CSF, which could imply a development defect at the level or near the arachnoid villi. Although the patient showed a severe internal hydrocephalus and a thinned cerebral parenchyma, we could not identify an area of possible cerebral parenchymal rupture on CT images.

Ultrasonography of the brain is a safe, minimally invasive modality to image the ventricles on young dogs through a persistent dorsal midline fontanelle.<sup>1</sup> Enlarged ventricles appear either as paired anechoic regions or, as in our case, as a single, large anechoic structure due to the absence of the *septum pellucidum* that normally separates the lateral ventricles. In human medicine, transfontanellar ultrasonography can easily make the diagnosis of benign external hydrocephalus by evaluating the skull-to-cortex distance that in children with this condition is greater than 5 mm.<sup>2</sup> In our case, the skull-to-cortex distance was of 8 mm; however, although no data are available regarding to the normal skull-to-cortex distance in dogs, it should be negligible as normal ultrasonography of intracranial structures does not reveal identifiable CSF in the subarachnoid space. CT can be considered as valuable tool for diagnosing hydrocephalus because it allows visualization of the entire ventricular system and subarachnoid space. CT findings indicative of internal hydrocephalus include

identification of distended lateral ventricles and increased ventricular volume.<sup>1</sup> In the case of external hydrocephalus, the hypoattenuating CSF, which is easily detected within the ventricular system, is seen in enlarged cortical sulci and the subarachnoid space.<sup>2</sup> However, the examination of choice in children with external hydrocephalus is MRI, which offers the best resolution and detailed evaluation of the brain parenchyma and is also more useful for identifying concurrent abnormalities.<sup>1,2</sup> Moreover, it appears essential in the differential diagnosis between benign enlargement of the subarachnoid spaces and subdural collections.

Findings in our case highlight that ultrasonography was a safe, minimally invasive modality to image the ventricles in the dog of the present report, which had a persistent fontanelle. However, more advanced imaging techniques should be performed following ultrasonographic evaluation if medical or surgical treatments are to be pursued.

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