A five-month-old female entire Chihuahua was referred to the neurology-neurosurgery service at Dick White Referrals for investigation of acute onset of fly catching episodes, left-sided head tilt and circling. General physical examination revealed presence of a large fontanelle, a dome-shaped head and papilloedema. On neurological evaluation, obtunded mental status, vestibular ataxia, bilaterally reduced menace response, spontaneous ventrolateral strabismus and increased segmental spinal cord reflexes were observed. The neuroanatomical localisation was consistent with a multifocal intracranial lesion. The suspected diagnosis based on signalment and clinical findings was congenital hydrocephalus; other differential diagnoses included various anomalous diseases, inflammatory/infectious and metabolic disorders.

Haematology and serum biochemistry were within normal limits. Magnetic resonance (MR; Hitachi Aperto open magnet 0.4T) of the brain was performed. T1-weighted, T2-weighted and fluid-attenuated inversion recovery (FLAIR) images were acquired in transverse, dorsal and sagittal planes. The images revealed severe distension of the lateral ventricles and atrophy of the cerebral cortex; slight dilation of the third ventricle was also present, while the ventricular system caudally to the mesencephalic aqueduct did not appear to be dilated. The mesencephalic aqueduct itself was subjectively small, definitely not distended when compared to the dilated lateral and third ventricles. Therefore, the obstruction to the cerebrospinal fluid (CSF) flow was located at the level of the mesencephalic aqueduct. MRI showed no evidence of cerebellar herniation, but a large open fontanelle, with brain parenchyma in the subcutaneous area, was observed. Findings were consistent with congenital hydrocephalus (Figures 1A and 1B).

Symptomatic therapy was initiated with Mannitol (0.5 g/kg once intravenously over 20 minutes), phenobarbitone (loading dose of 3 mg/kg four times daily initially, reduced to a maintenance dose of 3 mg/kg twice daily after 24 hours) and dexamethasone (0.1 mg/kg once daily initially, reduced to 0.1 mg/kg every other day after 48 hours). The dog improved remarkably and surgery for ventriculo-peritoneal shunt placement was performed.

The dog recovered well and was discharged seven days after surgery.
postoperatively, with improved neurological signs. Postoperative medications included cefuroxime (20 mg/kg intravenously, followed by cefalexin 22 mg/kg orally twice daily), dexamethasone (0.2 mg/kg intravenously, followed by prednisolone 1 mg/kg orally once daily, tapered to every other day four weeks later, and stopped completely five weeks postoperatively), phenobarbitone (3 mg/kg orally twice daily) and multivitamin supplementation. Perioperative analgesia was provided by methadone 0.3 mg/kg intramuscularly. The patient progressed to complete resolution of the clinical signs five months after surgery. At one year follow-up, no neurological deficits were present.

Hydrocephalus

The term hydrocephalus refers to the accumulation of excessive amounts of CSF within the cranial cavity, resulting in an abnormal dilation of the ventricular system or subarachnoid space (De Lahunta and Glass, 2009).

The two main categories of hydrocephalus are compensatory and obstructive (De Lahunta and Glass, 2009). In compensatory hydrocephalus, the volume of CSF increases to compensate for a loss or shrinkage of brain parenchyma following, for example, viral infections, ischaemic events or senile atrophy of the brain. Despite the increase in volume occupied by CSF, this type of hydrocephalus is characterised by a normal intracranial pressure (ICP). Conversely, obstructive hydrocephalus results from an obstruction to the CSF flow and absorption, with subsequent accumulation of fluid and concomitant increase in ICP. Causes of obstructive hydrocephalus can be classified as congenital, also called developmental, and acquired (Coates et al, 2006; De Lahunta and Glass, 2009). In veterinary patients, congenital forms of hydrocephalus are more common than acquired forms, with higher incidences reported in Maltese, Yorkshire Terrier, English Bulldog, Chihuahua, Lhasa Apso, Pomeranian, Toy Poodle, Cairn Terrier, Boston Terrier, Pug and Pekingese (Coates et al, 2006). Congenital obstructive hydrocephalus is suspected to be associated with stenosis of the mesencephalic aqueduct, resulting from a fusion of the rostral colliculi (Coates et al, 2006; De Lahunta and Glass, 2009). Malformations of the cerebellum and the caudal fossa have also been associated with congenital hydrocephalus. Abnormalities of the arachnoid villi have been hypothesised to be a cause in cases where a structural obstruction cannot be found. Acquired hydrocephalus results from neoplasia, inflammation, infection or degenerative disease interfering with CSF production, flow or absorption (Coates et al, 2006; De Lahunta and Glass, 2009).

Clinical signs and diagnosis

Type and severity of clinical signs are variable but forebrain signs tend to predominate (Coates, 2006; De Lahunta and Glass, 2009).

The lateral ventricles appear to be the most susceptible to an increase in CSF pressure and ventricular system expansion, with subsequent compression and functional compromise affecting mainly the diencephalon. However, some dogs display no neurological signs despite significant enlargement of the lateral ventricles and severe cerebral atrophy, suggesting that the clinical presentation may reflect the degree of CSF pressure (De Lahunta and Glass, 2009).

Clinical signs of congenital hydrocephalus are usually recognised within the first few months after birth; however, many animals do not develop clinical signs until later in life and are diagnosed as young adults (Shihab et al, 2011). Affected puppies are usually smaller than their littermates. Typical features of congenital hydrocephalus are an enlarged, dome-shaped calvarium.
and an open fontanelle. However, a persistent fontanelle is not always indicative of the presence or future development of hydrocephalus and many patients with congenital hydrocephalus may not have an open fontanelle. Bilateral ventrolateral strabismus is often detected on physical examination, and it is believed to be related to distortion of the orbit or brainstem dysfunction (Coates et al, 2006; Thomas, 2010). The main differential diagnoses when prosencephalic clinical signs are observed in an immature or young adult dog are hepatic encephalopathy associated with portosystemic shunts, infectious diseases and storage disease (De Lahunta and Glass, 2009).

Suspicion of hydrocephalus is based on signalment, history and physical examination. The diagnosis is usually confirmed with advance imaging (computed tomography and MR). If advance imaging is not immediately available, alternative diagnostic procedures can offer very useful information. Radiography (lateral projection of the skull) can provide evidence of distortion and thinning of the calvarium, presence of a persistent fontanelle, reduced prominence of the cerebral gyral pattern and a diffuse homogeneous appearance of the distended fluid-filled lateral ventricles. Ultrasound can be performed through an open fontanelle or the thin calvarial bone to visualise the lateral ventricles. CSF collection is associated with an increased risk of brain herniation and collapse of the cerebral parenchyma in cases of severe ventricular distension and increased ICP; CSF analysis should therefore only be performed after advanced imaging and careful evaluation of the individual case.

Treatment and management

Treatment can be medical or surgical. Medical treatment aims to reduce CSF production through the use of glucocorticoids and diuretics (Coates et al, 2006; Thomas, 2010). Omeprazole has been shown to decrease the CSF production in normal dogs but evidence of its effectiveness in patients with hydrocephalus is only anecdotal (Javaheri et al, 1997; Thomas, 2010).

Medical management offers only short-term control of the clinical signs, and surgical intervention is necessary in most symptomatic cases. Surgery is the treatment of choice in humans (Shihab et al, 2011); the procedure diverts CSF from the ventricles into another body cavity, most commonly the peritoneal cavity.

Surgical shunting

In animals, surgical shunting is recommended in cases presenting with severe neurological deficits and those refractory to medical therapy. Ventriculoperitoneal (VP) shunting is a palliative adjunctive treatment in animals with acquired hydrocephalus presenting severe clinical signs and when the underlying cause of the hydrocephalus cannot be eliminated, for example in cases of obstructive hydrocephalus caused by an unresectable intracranial mass (Coates et al, 2006; De Stefani et al, 2011). Contraindications to placing a VP shunt include increased CSF protein concentration, increased white blood cell or red blood cell count in the CSF, evidence of peritoneal inflammation or the presence of systemic infections, such as skin and urinary tract infections (Coates et al, 2006).

The shunt system consists of three basic components: a proximal ventricular catheter; a one-way pressure valve; and a distal peritoneal catheter. CSF-flow control valves operate at low, medium and high pressure; newer devices have adjustable pressure valves (Coates et al, 2006). During placement, the proximal catheter is positioned in the lateral ventricle through a burr hole created in the caudal aspect of the parietal bone and secured. The shunt is then tunnelled caudally in the subcutaneous tissue over the dorsolateral aspect of the cranium and the cervical region, so that the pressure valve is positioned at the level of the mid-cranial cervical spine. The distal portion of the shunting system is tunnelled caudally, the peritoneal catheter inserted into the abdominal cavity through a small flank approach caudal to the last rib and adequately secured. Shunting systems are usually made of radiopaque silicone to allow radiographic visualisation of the catheter and assess positioning (Figure 2) (Coates et al, 2006; De Stefani et al, 2011).

The most frequent complication associated with VP shunt placement is underdrainage with blockage of CSF flow due to...
collapse of brain parenchyma into the shunt, glosis, accumulation of blood or proteinaceous debris, scarring or adhesion around the distal catheter (Coates et al, 2006; Thomas, 2010; De Stefani et al, 2011). Misplacement, migration, kinking and disconnection of the shunting system components and infection may also occur (Thomas, 2010; De Stefani et al, 2011). Underdrainage usually manifests as a recurrence of the clinical signs; infection is accompanied by fever or signs of obstruction. Any of these complications usually require replacement of the shunt (Thomas, 2010). Another potential complication is functional failure from overdrainage (Coates, 2006). Overdrainage occurs as a consequence of excessive CSF flow from the ventricles, normally associated with the use of low-pressure valves in humans, and potentially resulting in subdural haematoma and cerebrocortical collapse (Platt et al, 2012); other signs reported in people are headache, vomiting, and changes in vision (Filgueiras Rda et al, 2009).

The failure rate of VP shunting in human patients varies between studies, but it is as high as 50% within the first two years postoperatively (Coates et al, 2006; Thomas, 2010). Similar complication rates have been reported in dogs, ranging from 21% to 70% (De Stefani et al, 2011; Platt et al, 2012). However, these figures may reflect the fact that these studies included various aetiologies for the hydrocephalus so considered an heterogeneous group of dogs, which makes this data potentially non-representative of a more homogeneous population. When a uniform population of dogs affected by congenital hydrocephalus is considered, as in the study by Shihab et al (2011), a complication rate of 25% is reported.

Prognosis is guarded to fair. In most cases, neurological signs improve soon after surgery; however, if deficits are severe, the postoperative improvement might be minimal, and permanent neurological abnormalities are likely if the cerebral cortex has been damaged (Thomas, 2010). It has been suggested that reconstitution of the cerebral hemispheres after shunting occurs only in the white matter, through a regeneration process characterised by myelin destruction, remyelination and reactive astrocytosis (Bagley, 2002; Coates et al, 2006; Kitagawa et al, 2008).

Conclusion
Despite risks and potential complications, shunting is still considered the best option for many patients with hydrocephalus, and the only treatment able to offer long-term symptom control.

Good communication with owners is paramount when discussing the pros, cons and implications of VP shunting. A realistic expectation is an improvement in neurological signs, not a complete resolution (Coates et al, 2006). In a recent study (De Stefani, 2011), persistence of seizures postoperatively was the most common reason for euthanasia; this underlines the importance of discussing expectations with clients. For a good outcome, the owner’s commitment is essential; most medications need to be administered several times a day and the shunt requires lifelong care.

Shunting is the treatment of choice for hydrocephalus in humans. Many veterinary patients experience a significant improvement in quality of life after shunting, with a level of persistent neurological abnormalities that is considered acceptable. A successful outcome is more likely in animals with minimal clinical signs as damage to cortical neurons is irreversible; this shows the importance of treating any case of hydrocephalus promptly and aggressively (Bagley, 2002; 2004).

References

KEY POINTS
- Congenital hydrocephalus often manifests with forebrain deficits
- Surgery to place a shunt to control CSF flow is the treatment of choice, despite risks and potential complications
- Shunting is more likely to improve than resolve symptoms

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Adjunctive tests

1: Which is not an indication for VP shunt placement?
   a. Congenital obstructive hydrocephalus
   b. Compensatory hydrocephalus
   c. Severe neurological deficits or refractory to medical therapy
   d. Acquired obstructive hydrocephalus

2: Which statement on medical treatment of hydrocephalus is incorrect?
   a. It involves using mainly glucocorticoids and diuretics
   b. It aims to reduce the CSF production
   c. It offers short-term control of the clinical signs
   d. It is the treatment of choice in many hydrocephalus cases

3: What is the most frequent complication associated with VP shunt placement?
   a. Underdrainage
   b. Overdrainage
   c. Infection
   d. Disconnection of the shunting system components

For answers please see page 242