A REPRINT FROM

COMPENDIUM ON CONTINUING EDUCATION FOR THE PRACTICING VETERINARIAN

Continuing Education Article #7

Vol. 13, No. 2, February 1991 245

Congenital Deafness in Dogs and Cats

KEY FACTS

- Deafness may result from outer and middle ear abnormalities (i.e., conductive deafness) or from cochlear abnormalities (i.e., sensorineural deafness). Although conductive deafness is rare, it may be amenable to surgical repair; sensorineural deafness is not treatable.
- Sensorineural deafness may result from neuroepithelial degeneration, in which the primary event is cochlear hair cell loss, or from cochleosaccular degeneration, in which hair cell loss is secondary to degeneration of the stria vascularis.
- Certain animal breeds, such as blue-eyed white cats, dalmatians, English setters, and Australian shepherds, may be especially prone to congenital deafness but the deficit may appear in any breed.
- Congenital deafness is frequently associated with the piebald gene, the merle gene, or pigmentation abnormalities and may be part of a spectrum of abnormalities, including heterochromia iridis (blue irides), absence of retinal pigmentation, and partial albinism (piebaldism).
- Diagnosis of unilateral deafness requires brain stem auditory-evoked potential testing at referral centers, while bilateral deafness can generally be evaluated in the clinic.

Louisiana State University
George M. Strain, PhD

THE CLINICIAN faced with an owner's complaint of deafness in a pet is usually in a difficult position. Unilateral deafness is difficult to detect, vibrations from typical clinical auditory test stimuli may be detected by tactile somatosensory receptors, and stoic or refractory animals may be unresponsive. Further, questions about future breedings of affected animals may be hard to answer. In this article, the current state of knowledge about congenital deafness in dogs and cats is reviewed and diagnostic aids are described.

PHYSIOLOGY AND ANATOMY OF HEARING

When air molecule vibrations of sound reach the outer ear, they are conducted into the external auditory canal where frequencies of importance to the species may be amplified by the tuning characteristics of the canal. The shape of the external ear structures acts as a collector for sound and is useful in sound localization. Air vibrations in the auditory canal produce sympathetic vibrations in the taut tympanic membrane; sympathetic vibrations in turn are transmitted through a series of three bones to the oval window of the cochlea located in the petrous portion of the temporal bone (Figure 1). Vibrations of the tympanic membrane, which may be as small as the diameter of a hydrogen molecule, are transmitted with negligible distortion but with increased power by the auditory ossicles of the middle ear: the malleus, the incus, and the stapes. Damage to these structures can result in deafness (conductive) despite a normal cochlea. Muscle tension in the tensor tympanii muscle attached to the tympanic membrane and the stapedius muscle (the smallest striated muscle in the body) attached to the stapes allows the animal to reduce or enhance sensitivity (e.g., the cochlea is not overloaded while a dog barks).[1]

Vibrations transmitted through the ossicles to the oval window result in vibrations of perilymph (fluid) of the scala vestibuli (Figure 2). This canal joins the scala tympani at the apex (helicotrema) of the cochlea so that inward deflections of the oval window produced by the stapes result in an outward deflection of the round window; this relief mechanism is necessary because the fluids involved are relatively incompressible and the entire cochlea is en-

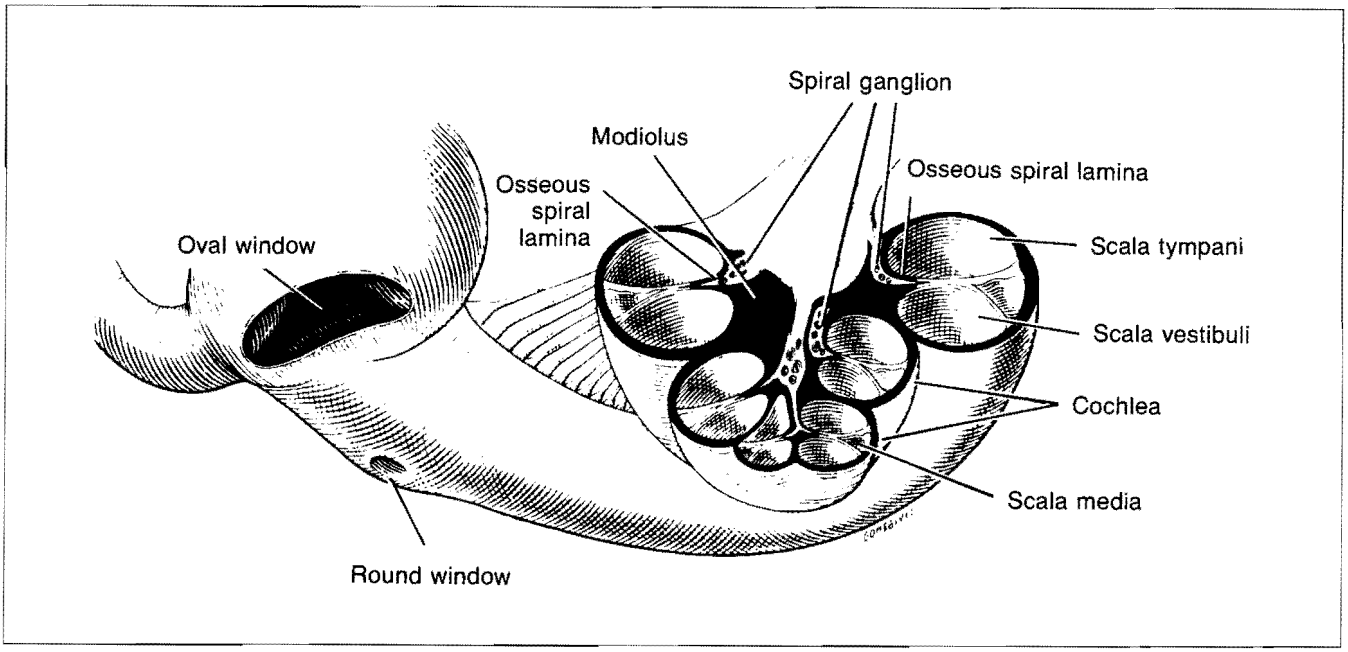

Figure 1—Schematic diagram of the cochlea. Vibrations transmitted to the oval window by the stapes (not shown) result in out-of-phase vibrations at the round window. (From Junqueira LC, Carneiro J: *Basic Histology*, ed 4. Los Altos, CA, Lange Medical Publications, 1983. Reproduced with permission.)

cased in unyielding bone. Inward movement of fluid in the scala vestibuli and compensatory outward movement in the scala tympani result in a shearing force being applied to the tectorial membrane of the cochlear duct. Within the cochlear duct, organ of Corti hair cell stereocilia are embedded in the tectorial membrane so that shearing forces from sound waves propagated down the scala vestibuli produce bending of the stereocilia, thereby depolarizing their cells. Because of the physical nature of the basilar membrane and the waves themselves, high frequencies are most effective in activating hair cells near the base of the cochlea while low frequencies best activate hair cells near the apex. Synaptic connections between hair cells and spiral ganglion neurons result in communication of information about hair cell activity through the eighth cranial nerve to the central nervous system. The stria vascularis lining the outer wall of the cochlear duct is the major vascular area of the cochlea and produces endolymph, which is necessary for maintenance of cochlear hair cells.[2,3] Sensorineural deafness may result from direct damage to the hair cells or from hair cell loss secondary to strial degeneration.

Auditory function is present when the ear canals open at age 12 to 14 days in dogs and five days in cats, although behavioral responses to loud noises can be detected before the canal opens.[4] Reported hearing ranges are from 67 Hz to 45 kHz in dogs and 45 Hz to greater than 64 kHz in cats, while the range for humans is 64 Hz to 23 kHz.[5-7] When based on brain stem auditory-evoked potential measurements, hearing threshold reaches the most sensitive level by 20 days of age in dogs[8] and 30 days of age in cats.[9]

INCIDENCE AND CAUSE

The incidence of congenital deafness in the general canine and feline population is unknown, but the occurrence of deafness may reach 30% in selected breeds. A retrospective search of records from 1.1 million canine visits to 14 U.S. veterinary medical teaching hospitals during a 14-year period[10] revealed an incidence of 2.56 cases per 10,000; while a survey in Australia found 12 canine cases during a one-year period in reports from 37 veterinary practices[11] or an incidence of 0.32 cases per practice per year. Because these reports reflect only dogs presented for medical evaluation, and because unilateral deafness is more common and more difficult to detect than bilateral deafness is, and because breeders frequently euthanatize deaf puppies, the true incidence of deafness in dogs is certainly much higher. Less is known of the incidence of deafness in cats, for which the incidence of congenital defects is said to be the lowest of all domestic animals.[12] In humans, one in every 750 children is congenitally deaf, with at least half of the cases attributable to genetic disorders.[13]

CANINE BREEDS for which congenital deafness has been reported are listed on page 248.[14-21] Practitioners should realize, however, that any breed of dog (or cat) can

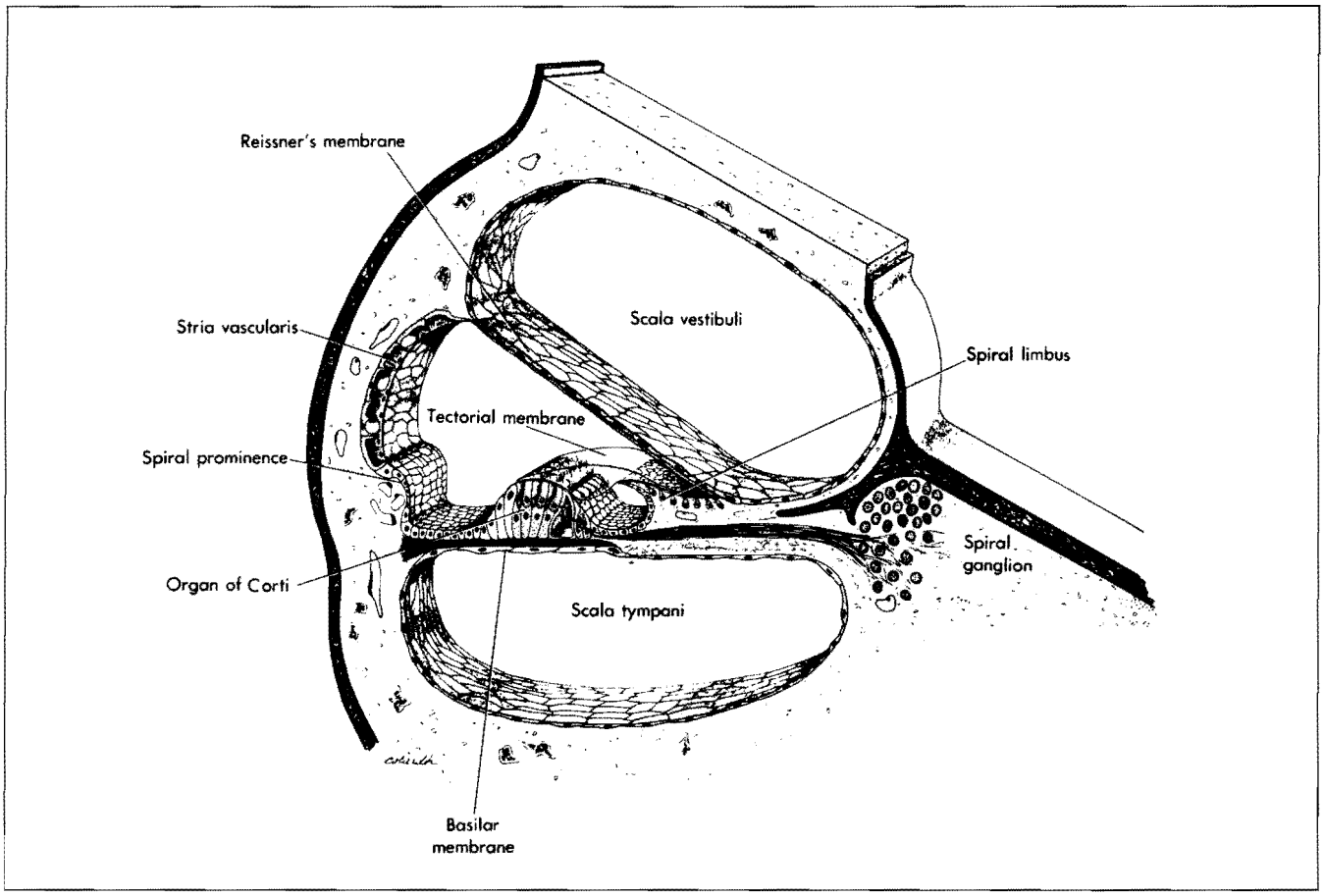

Figure 2—Schematic diagram of a cross section through one of the turns of the cochlea. The organ of Corti is located in the cochlear duct formed by the basilar membrane and Reissner's membrane. Organ of Corti hair cell stereocilia are embedded in the tectorial membrane so that sound waves propagated down the scala vestibuli produce shearing of the stereocilia, thereby depolarizing their cells. The stria vascularis lining the outer wall of the cochlear duct produces endolymph, which is necessary for hair cell maintenance. (From Bloom W, Fawcett DW: *A Textbook of Histology*, ed 10. Philadelphia, WB Saunders Co, 1975. Reproduced with permission.)

potentially be affected. The deafness in many canine breeds has been shown to be hereditary. Reports of congenital deafness in cats are not available by breed, but it is widely recognized that blue-eyed white cats have a high incidence of deafness.[22] Little information about the incidence of congenital deafness by breed is available, however. In one survey, the highest risk values were shown for dalmatians, followed by Australian heelers, English setters, and Australian shepherds.[10] The incidence for dalmatians has been shown to be 8% bilateral deafness and 22% unilateral deafness for a total of 30% of all dogs.[23,24] Unilateral or bilateral deafness affects 75% of all white Norwegian dunkerhounds,[25] but the incidence in normal-color dogs is unknown. White cats constitute approximately 5.7% of the overall population, and 15% of these are blue in one or both eyes.[16] Because there is a positive correlation between blue eyes and deafness in cats,[26] this statistic provides a crude estimate of a 1% incidence of deafness in white cats. The number of hearing blue-eyed white cats is approximately offset by deaf white cats without blue eyes.

The causes of congenital deafness can be separated into acquired and hereditary (see the listing on page 248). Acquired deafness results from prenatal and perinatal events, such as malformations, viral infection, or transplacental exposure to ototoxic compounds.[1,13,19] Although correction of the deafness in most of these cases is not possible when the cause is correctly identified, the animals may be safely bred without fear of deaf offspring. The incidence of deafness from these causes is probably negligible in dogs and cats, although it may account for half of the human cases. Hereditary deafness may result from autosomal dominant or recessive transmission or may be sex-linked. Most genetic transmission of canine and feline deafness evidently results from autosomal dominant genes, although at least one recessive canine syndrome has been described (bullterrier[27]). Sex-linked hereditary deafness has not been documented in dogs or cats, although it accounts for 3% of congenitally deaf humans.[28] Breeding of animals with

Canine Breeds with Reported Congenital Deafness[14-21]

Akita	Fox terrier
American Staffordshire terrier	Great Dane
Australian heeler	Great Pyrenees
Australian shepherd	Maltese
Beagle	Miniature poodle
Border collie	Mongrel
Boston terrier	Norwegian dunkerhound
Bullterrier	Old English sheepdog
Catahoula leopard dog	Papillon
Cocker spaniel	Pointer
Collie	Rhodesian ridgeback
Dalmatian	Scottish terrier
Dappled dachshund	Sealyham terrier
Doberman pinscher	Shetland sheepdog
Dogo Argentino	Shropshire terrier
English bulldog	Walker American foxhound
English setter	West Highland white terrier
Foxhound	

Causes of Congenital Deafness

Acquired	Hereditary
Meningitis	Autosomal dominant
Viral and other infections	Autosomal recessive
Kernicterus	X-linked
Anoxia	Unknown
Otitis	
Ototoxicity	
Noise	
Malformations	
Unknown	

hereditary deafness will generally ensure a greater than normal incidence of deaf offspring as well as propagation of an undesirable defect in the gene pool of the breed.[23]

CLINICAL SIGNS

Assessment of auditory function cannot be performed before the ear canals open (12 to 14 days of age in dogs and five days of age in cats) because of the sound-insulating properties of sealed ear canals. After affected puppies and kittens reach their respective ages, they show a spectrum of behavior highly suggestive of bilateral deafness. Deaf animals are usually difficult to arouse from sleep at feeding time and only respond to tactile stimuli. They are usually more aggressive with littermates because they do not hear the pain vocalizations that normally terminate such behavior in hearing animals. In general, deaf animals also are extremely vocal when separated from littermates because they become highly dependent on the physical presence of littermates.

Owners of a deaf dog or cat may not become suspicious of the deficit until the pet reaches one year of age or older, previously attributing the behavior to stubbornness or stupidity. This tendency is particularly true when deafness is unilateral. Such animals demonstrate difficulty in localizing sound sources in the absence of normal binaural cues and may be difficult to awaken when sleeping in lateral recumbency on the healthy ear.

Breeders or owners of deaf puppies or kittens are best advised to euthanatize bilaterally deaf animals because care of these pets is difficult and frequently they die early from vehicular accidents or other unperceived danger. For example, bilaterally deaf animals that are startled while asleep may reflexly bite; furthermore, they will not respond to voice commands to go outside for bowel or bladder relief. It is possible, however, to raise deaf animals, especially housebound pets, successfully. Some owners train pets to respond to a flash of the porch light, and it has been suggested that aversive-training collars that electrically shock the skin can be used at the lowest setting to call deaf dogs. A highly dedicated owner, however, is required to care for these animals.

DIAGNOSTIC PROCEDURES

Evaluation of auditory function is usually performed by producing a sound outside the animal's visual field or while the animal is blindfolded plus observation of such responses as Preyer's reflex (pinna movement). This dual approach can be useful for bilaterally deaf animals, although caution must be exercised to prevent the sound from generating vibrations through the floor or a table, as dogs and cats can detect such vibrations through the somatosensory system. Appropriate cues include jingling of keys, whistles, or hand claps. Unfortunately, highly stressed animals with intact hearing may not respond and deaf animals may quickly sense the presence of the unseen tester. Unilaterally deaf animals can be evaluated in a similar manner; that is, by observing for efforts at sound localization while the stimulus is produced in different areas of the room. The results are usually equivocal, however.

Conclusive diagnosis of unilateral or bilateral deafness requires brain stem auditory-evoked potential (BAEP) testing, which is usually available at schools of veterinary medicine and some referral centers. With this test, potentials generated in the cochlea and brain stem by auditory stimuli are detected with special-purpose instrumentation.[8] A highly repeatable series of five or six peaks is present in the response, each is assigned a Roman numeral, and the latencies from stimulus onset to each peak are known for normal animals. The test does not require cooperation on

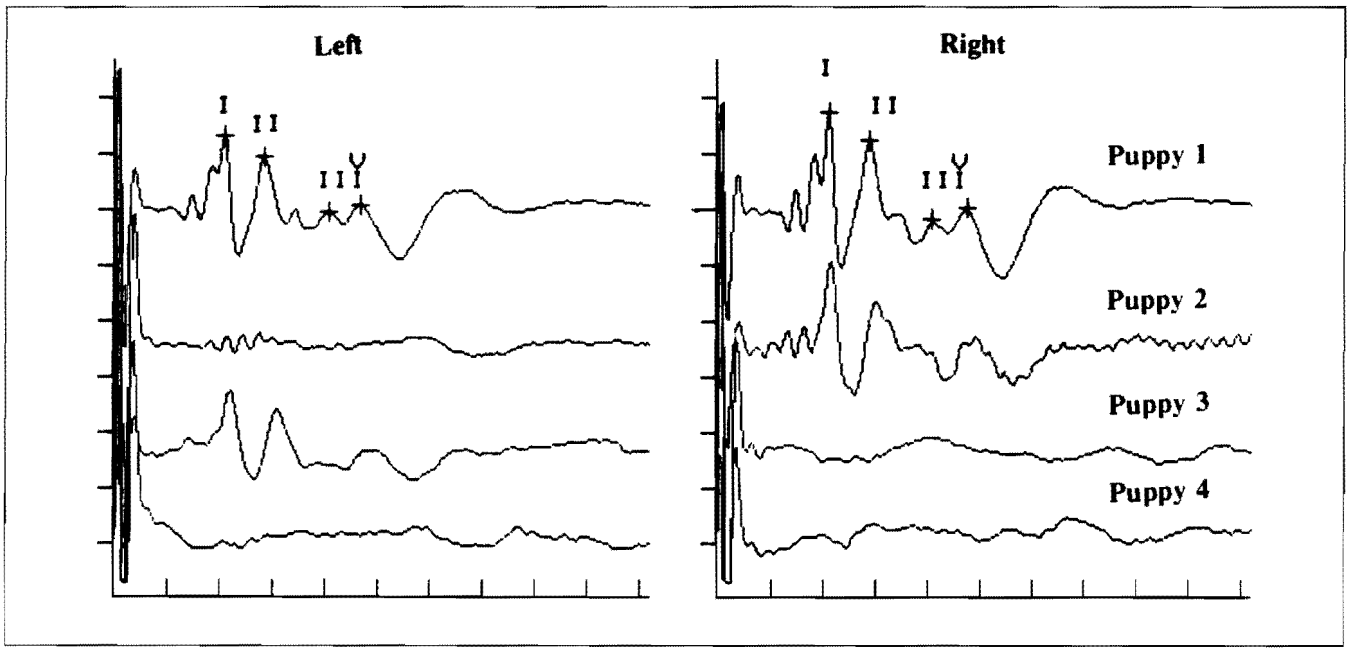

Figure 3—Brain stem auditory-evoked potentials recorded from four dalmatian puppies demonstrate responses in normal (top), unilaterally deaf (middle two), and bilaterally deaf (bottom) animals. Evidence of minor residual cochlear function is apparent in the left ear response of the first unilateral puppy, occurring at the normal latency for peak I of the response. (Horizontal axis: 1 ms/div; vertical axis: 1.5 μV/div)

the part of the subject and can usually be performed without chemical restraint. The first peak in the response, peak I, is generated in the cochlea so that cochlear deafness is demonstrated by the absence of that peak and all subsequent peaks. Brain stem auditory-evoked potentials are present when the ear canals open and reach mature patterns by 30 to 40 days of age in dogs[8] and three to four weeks of age in cats.[9] The potentials recorded from four five-week-old dalmatians are shown in Figure 3. The normal responses for both ears are shown on top, followed by two unilaterally deaf puppies and one deaf puppy. A suggestion of minor residual cochlear function is apparent in the left ear response of Puppy 2. At Louisiana State University, hearing screening is not usually performed on puppies until five weeks of age, when cochlear degeneration is sufficiently complete (at least in dalmatians) to produce results. Dalmatian breeders, and more recently English setter breeders, are actively promoting auditory testing of litters in an effort to lower the incidence of deafness. Practicing veterinarians should be aware of this trend and should refer owners of these breeds to appropriate testing facilities.

PATHOPHYSIOLOGY

Peripheral deafness may result from outer and middle ear abnormalities (conductive deafness), such as tympanic membrane defects and ossicle fusion (rare but may be amenable to surgical repair) or from cochlear abnormalities (sensorineural deafness). (Even more rare is deafness from morphogenetic causes in which inner ear structural development is disturbed in early embryonic stages.) Central deafness, which is also rare, results from retrocochlear lesions. Cochlear sources of deafness may in turn be divided into cochleosaccular (Scheibe) degeneration and neuroepithelial degeneration.[29] Most hereditary deafness results from the former, while acquired deafness usually results from the latter. In cases of neuroepithelial degeneration, the primary abnormality is in the organ of Corti, with the Reissner's membrane intact and the stria vascularis initially normal. In mouse mutants of this disorder, the organ of Corti is nearly mature before degeneration and outer hair cells degenerate before inner hair cells.[3] In cases of cochleosaccular degeneration, the disorder is vascular, with the stria vascularis affected first, followed by collapse of Reissner's membrane and degeneration of the organ of Corti. Normal pigment cells, which apparently are necessary for normal function, are absent in the stria.[30,31] Most early studies of cochleosaccular degeneration were performed on dalmatians[32-38] and white cats,[22,36] although several mouse mutants are now widely used.[3]

DEAFNESS in dogs is frequently associated with the presence of the merle (M) gene (collie, Shetland sheepdog, harlequin Great Dane, dappled dachshund, American foxhound, Norwegian dunkerhound) and the piebald (S^P) or

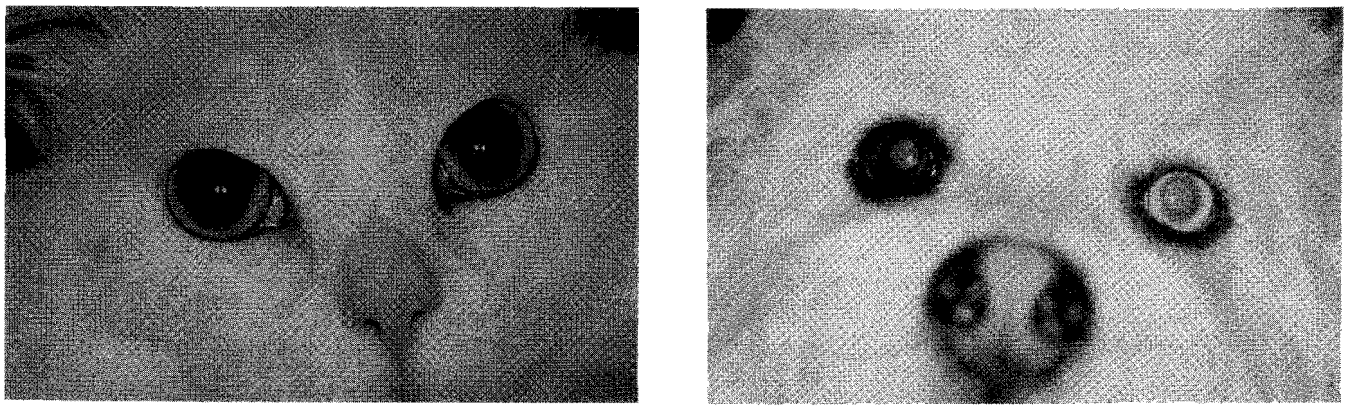

Figure 4—(**A**) A cat with bilateral heterchromia iridis and (**B**) a dog with unilateral heterochromia iridis (absence of pigmentation in the iris or irides). Red reflected from the back of the retina occurs because of the frequent association of heterochromia iridis with absence of tapetal pigmentation; the red color results from remaining blood vessels. Neither animal is deaf. (Photograph of dog courtesy of M. B. Glaze, DVM, MS, Louisiana State University)

extreme piebald (S^w) gene (bullterrier, Samoyed, Great Pyrenees, Sealyham terrier, greyhound, bulldog, dalmatian, beagle),[39] where large amounts of white are present in the fur; however, deafness has not been reported for all of these breeds. Hereditary deafness associated with pigmentation abnormalities has been reported as a syndrome with a spectrum of features, including piebaldism or partial albinism, deafness, heterochromia iridis (blue iris or irides attributable to absence of pigment, as shown in Figure 4), absence of retinal pigment, absence of cochlear stria vascularis pigment, and various facial defects.[26] The syndrome is usually inherited as an autosomal dominant trait with incomplete penetrance so that affected individuals may not display all components. The syndrome is seen in humans (Waardenburg's syndrome), cats, dogs, mink, mice,[26] cattle,[40] and horses,[41] although the deafness has not been investigated in cows or horses and the mode of inheritance has not clearly been established for all species or breeds (e.g., dalmatian). The critical factor in determining deafness apparently is the absence of melanocytes in the stria vascularis; these cells perform some as yet unknown function in this structure, which in turn is necessary for maintenance of cochlear hair cells.[29,30] Strial melanocytes and other affected tissue in animals with this syndrome have been shown by embryologic studies to originate in neural crest cells.[42]

CONCLUSION

Congenital deafness may be frequently seen in blue-eyed white cats and in dalmatians, English setters, and Australian shepherd breeds, among others, but is generally uncommon in the overall canine and feline populations. The deafness may be either acquired or hereditary, although the latter is seen most often. Conductive hearing loss attributable to outer or middle ear abnormalities is less common than is sensorineural deafness involving the inner ear. Sensorineural deafness may result from neuroepithelial degeneration, in which hair cells are primarily affected, or from cochleosaccular degeneration, in which hair cell loss is secondary to degeneration of the stria vascularis. Definitive diagnosis of unilateral or bilateral deafness requires brain stem auditory-evoked potential testing, although bilateral deafness can usually be detected by simple office testing.

About the Author

Dr. Strain is Associate Professor of Neuroscience for the Department of Veterinary Physiology, Pharmacology and Toxicology, and Adjunct Associate Professor in the Department of Communication Disorders, School of Veterinary Medicine, Louisiana State University, Baton Rouge, Louisiana.

REFERENCES

1. Myers D, Schlosser WD, Wolfson RJ, et al: Otologic diagnosis and the treatment of deafness. *Clin Symp* 22:34–69, 1970.
2. Nachlas NE, Lurie MH: The stria vascularis: Review and observations. *Laryngoscope* 61:989–1003, 1951.
3. Harrison RV: *The Biology of Hearing and Deafness*. Springfield, IL, CC Thomas, 1988, pp 25–26.
4. Foss I, Flottorp G: A comparative study of the development of hearing and vision in various species commonly used in experiments. *Acta Otolaryng* 77:202–214, 1974.
5. Heffner HE: Hearing in large and small dogs: Absolute thresholds and size of the tympanic membrane. *Behav Neurosci* 2:310–318, 1983.
6. Heffner RS, Heffner HE: Hearing range of the domestic cat. *Hear Res* 19:85–88, 1985.
7. Fay RR: *Hearing in Vertebrates: A Psychophysics Databook*. Winnetka, IL, Hill-Fay Associates, 1988.
8. Strain GM, Tedford BL, Jackson RM: Postnatal development of the brainstem auditory-evoked potential in dogs. *Am J Vet Res*, in press.
9. Buchwald JS, Shipley C: Development of auditory evoked potentials in the kitten, in Aslin RN (ed): *Advances in Neural and Behavioral Development*, vol 2. Norwood, NJ, Ablex Publishing Corp, 1986, pp 95–118.
10. Hayes HM, Wilson GP, Fenner WR, et al: Canine congenital deafness: Epidemiologic study of 272 cases. *JAAHA* 17:473–476, 1981.
11. Johnston DE, Cox B: The incidence in purebred dogs in Australia of abnormalities that may be inherited. *Austr Vet J* 46:465–474, 1970.
12. Saperstein G, Harris S, Leipold HW: Congenital defects in domestic cats. *Feline Pract* 6(4):18–43, 1976.

13. Epstein S, Reilly JS: Sensorineural hearing loss. *Ped Clin North Am* 36:1501–1520, 1989.
14. Erickson F, Saperstein G, Leipold HW, McKinley J: Congenital defects of dogs. Part 1. *Canine Pract* 4(4):54–61, 1977.
15. Oliver JE Jr, Hoerlein BF, Mayhew IG: *Veterinary Neurology*. Philadelphia, WB Saunders, 1987, p 198.
16. Braund KG: *Clinical Syndromes in Veterinary Neurology*. Baltimore, Williams & Wilkins, 1986, pp 3–5.
17. Hoerlein BF: *Canine Neurology*, ed 3. Philadelphia, WB Saunders, 1978, p 370.
18. deLahunta A: *Veterinary Neuroanatomy and Clinical Neurology*, ed 2. Philadelphia, WB Saunders, 1983, pp 308–309.
19. Neer TM: The ear, in Hoskins JD (ed): *Veterinary Pediatrics*. Philadelphia, WB Saunders, 1990, pp 459–472.
20. Clark RD, Stainer JR: *Medical & Genetic Aspects of Purebred Dogs*. Edwardsville, KS, Veterinary Medicine Publishing Co, 1983, pp 475–552.
21. Strain GM: Unpublished observations (American Staffordshire terrier, Catahoula, papillon, Dogo Argentino, Rhodesian ridgeback), Louisiana State University, 1990.
22. Mair IWS: Hereditary deafness in the white cat. *Acta Otolaryngol [Suppl]* 314:1–48, 1973.
23. Strain GM, Kearney MT, Gignac IJ, et al: Brainstem auditory evoked potential assessment of congenital deafness in dalmatians: Correlations with phenotypic markers. *J Vet Internal Med*, in press, vol 5, 1991.
24. Holliday TA, Nelson HJ, Williams DC, et al: Incidence of unilateral and bilateral brainstem auditory evoked response abnormalities in 900 dalmatian dogs. *Proc 7th Annu Vet Med Forum (ACVIM)* 7:1048, 1989.
25. Foss I: Development of hearing and vision, and morphological examination of the inner ear in hereditary deaf white Norwegian dunkerhound and normal dogs (black and dappled Norwegian dunkerhounds). MS Thesis, Cornell University, 1981.
26. Brown KS, Bergsma DR, Barrow MV: Animal models of pigment and hearing abnormalities in man, in Bergsma D (ed): *Birth Defects: Original Article Series*, vol VII, no 4. Baltimore, Williams & Wilkins, 1971, pp 102–109.
27. Erickson F, Saperstein G, Leipold HW, McKinley J: Congenital defects of dogs. Part 2. *Canine Pract* 4(5):51–61, 1977.
28. Gerber SE, Mencher GT: *Auditory Dysfunction*. Houston, College-Hill Press, 1980, pp 83–109.
29. Steel KP, Barkway C, Bock GR: Strial dysfunction in mice with cochleo-saccular abnormalities. *Hear Res* 27:11–26, 1987.
30. Schrott A, Spoendlin H: Pigment anomaly-associated inner ear deafness. *Acta Otolaryngol* 103:451–457, 1987.
31. Savin C: The blood vessels and pigmentary cells of the inner ear. *Ann Otol* 74:611–622, 1965.
32. Rawitz B: Gehororgan und Gehirn eines weissen Hundes mit blauen Augen. *Morpholog Arbeiten* 6:545–553, 1896.
33. Lurie MH: The membranous labyrinth in the congenitally deaf collie and dalmatian dog. *Laryngoscope* 58:279–287, 1948.
34. Hudson WR, Ruben RJ: Hereditary deafness in the dalmatian dog. *Arch Otolaryngol* 75:213–219, 1962.
35. Anderson H, Henricson B, Lundquist P-G, et al: Genetic hearing impairment in the dalmatian dog. *Acta Oto-Laryngol [Suppl]* 232:1–34, 1968.
36. Suga F, Hattler KW: Physiological and histopathological correlates of hereditary deafness in animals. *Laryngoscope* 80:81–104, 1970.
37. Johnsson L-G, Hawkins JE Jr, Muraski AA, et al: Vascular anatomy and pathology of the cochlea in dalmatian dogs, in de Lorenzo AJD (ed): *Vascular Disorders and Hearing Defects*. Baltimore, University Park Press, 1973, pp 249–295.
38. Mair IWS: Hereditary deafness in the dalmatian dog. *Arch Otorhinolaryngol* 212:1–14, 1976.
39. Little CC: *The Inheritance of Coat Color in Dogs*. New York, Howell Book House, 1957.
40. Leipold HW, Huston K: A herd of glass-eyed albino hereford cattle. *J Hered* 57:179–182, 1966.
41. Mayhew IG: *Large Animal Neurology*. Philadelphia, Lea & Febiger, 1989, p 195.
42. Weston JA: The migration and differentiation of neural crest cells. *Advanc Morphogen* 8:41–114, 1969.

ARTICLE #7 REVIEW QUESTIONS

The article you have read qualifies for ½ hour of Continuing Education Credit from the University of Pennsylvania School of Veterinary Medicine. *Choose only the one best answer* to each of the following questions; then mark your answers on the registration form inserted in *The Compendium.*

1. The initial event in cochleosaccular sensorineural deafness is degeneration of the
   a. saccule.
   b. stria vascularis.
   c. inner hair cells.
   d. outer hair cells.
   e. Reissner's membrane.

2. In the pigment abnormality–associated syndrome that includes human Waardenburg's syndrome and blue-eyed white cats, deafness is frequently associated with
   a. autosomal recessive transmission.
   b. urogenital defects.
   c. absence of retinal pigmentation.
   d. ventral septal defects.

3. Inward deflection of the oval window resulting from sound is accompanied by
   a. shearing of hair cell stereocilia by Reissner's membrane.
   b. outward deflection of the round window.
   c. vestibular stimulation.
   d. outward movement of the stapes.

4. Diagnosis of deafness by brain stem auditory-evoked potential (BAEP) testing
   a. shows a series of 10 peaks labeled I through X in normal-responding ears.
   b. requires sedation of the subject.
   c. shows only one peak (peak I) in deaf ears.
   d. shows a flat line response in deaf ears.

5. Acquired deafness may result from all of the following except
   a. anoxia.
   b. malformation.
   c. noise.
   d. meningitis.
   e. tetralogy of Fallot.

6. The behavior of deaf puppies is usually distinguished by
   a. passivity.
   b. reduced appetite.
   c. photophobia.
   d. aggressive play with littermates.
   e. independence.

7. Opening of the ear canals
   a. occurs by 12 to 14 days of age in puppies and kittens.
   b. is the time when hearing is first present.
   c. is the time when hearing testing is first practical.
   d. is usually delayed in congenitally deaf animals.

8. Cochlear structures are derived embryologically from the
   a. neural crest.
   b. notochord.
   c. neural tube.
   d. mesoderm.
   e. entoderm.

9. Deafness in dogs is frequently associated with the gene for
   a. merle (M) and extreme piebald (s^w).
   b. ticking (t).
   c. brindle (e^{br}).
   d. agouti or wild-color (a^w).
   e. lightening (g).

10. Cochlear hair cells
    a. are located on Reissner's membrane.
    b. are sensory receptors of primary sensory neurons whose cell bodies are located in the spiral ganglia.
    c. located in the basilar portion of the cochlea respond best to low-frequency sounds.
    d. are damaged in animals with conductive deafness.
    e. are depolarized by bending of their stereocilia.