AETIOLOGY, PREVALENCE AND DIAGNOSIS OF DEAFNESS IN DOGS AND CATS

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SUMMARY

Peripheral deafness may be inherited or acquired, congenital or later-onset, and sensorineural or conductive. The most commonly observed forms are inherited congenital sensorineural, acquired later-onset sensorineural (ototoxicity, presbycusis) and acquired later-onset conductive (chronic otitis externa/media). In most dog and cat breeds inherited congenital sensorineural deafness results from perinatal degeneration of the stria vascularis, the vascular bed of the outer wall of the cochlear duct, which leads to hair cell degeneration. The strial degeneration appears to result from the absence of melanocytes, but their function in this structure is unknown. Ototoxicity may result from any of a large number of drugs and chemicals that directly or indirectly destroy cochlear hair cells. The effects are dose-dependent and in rare cases reversible. The most commonly recognized ototoxic drugs are the aminoglycoside antibiotics. Presbycusis, the ageing-related progressive hearing loss unattributable to other causes, is sensorineural but may also include mechanical changes in the tympanum and ossicles. Hearing aids may be accepted by some dogs as long as some residual function remains. Breeds reported to have been affected by congenital sensorineural deafness are listed and those with the highest prevalence are noted. Methods for diagnosis of deafness are described.

KEYWORDS: Deafness; ototoxicity; presbycusis; brain-stem auditory evoked potential; Dalmation.

INTRODUCTION

An animal without auditory function is at a disadvantage that can range from trivial to extreme. The dog or cat with unilateral deafness experiences difficulty localizing the source of sound but quickly learns to compensate. However, a
bilaterally deaf animal is unable to anticipate dangers such as motor vehicles or predators and may, as a result, fall victim to injury or death. Diminished auditory function can likewise be merely inconvenient or hazardous. The causes of hearing loss and deafness are varied, and the implications for management and future breeding vary accordingly.

AUDITORY STRUCTURE AND FUNCTION

The auditory system may be considered to consist of three components (Evans & Christensen, 1979): the outer, middle and inner ears. The outer ear, consisting of the pinna and ear canal extending up to the tympanic membrane, serves to direct sound waves toward the receptor organ. Considerable variation exists in the conformation of the pinna between species and breeds, but the attached muscles enable orientation of the external ear toward sound sources (King, 1993). Coordinated movements of both pinnae still occur in animals with unilateral deafness when alerting to auditory stimuli. Analyses of the human ear canal have shown that its shape and dimensions optimize the transmission of the sound frequencies important in speech communications, but similar analyses have not been done for domestic species. Apocrine glands in the skin of the canal produce cerumen, a sebaceous product of cellular breakdown that serves to cleanse the canal (Johnson & Hawke, 1988). Great variation in the rate of cerumen production occurs, with long-haired dog breeds being more prolific cerumen producers and hence requiring greater grooming attention. Chronic infections may result in stenosis or occlusion of the canal and blockage of sound transmission.

The air-filled middle ear includes the tympanic membrane, the ossicles (malleus, incus and stapes), their associated muscles and ligaments and the opening of the auditory tube, which provides communication with the pharynx as well as a route for infection. Sound vibrations in the ear canal are transmitted to the tympanic membrane, and in turn are transmitted through the articulations of the ossicles to the attachment of the foot plate of the stapes on the membrane of the oval window. The ossicles amplify the vibrations of sound and in turn pass them on to the fluid-filled inner ear. The ossicular muscles, the stapedius and the tensor tympani, enable reflex damping of sound transmission in response to abrupt noises and in anticipation of loud vocalization by reducing ossicle movement. Innervation of the stapedius and tensor tympani muscles is by the trigeminal and facial nerves, respectively.

The cochlea (Latin, meaning snail shell) and the semicircular canals constitute the inner ear. The cochlea, a coiled structure enclosing three fluid-filled chambers (Fig. 1), is encased in the temporal bone with two membranous surfaces exposed at its base: the oval window and the round window. The foot plate of the stapes adheres to the oval window, transmitting sound vibrations into the cochlea. Two of the three cochlear chambers are contiguous at the apex. Inward deflections of the oval window caused by the foot plate of the stapes compress the fluid in the scala vestibuli; this compression wave travels along the coils of the cochlea in the scala vestibuli to the apex, then travels back down the coils in the scala tym-
The round window serves as a pressure-relief vent, bulging outward with inward deflections of the oval window. The third cochlear chamber, the scala media or cochlear duct, is positioned between the scala vestibuli and scala tympani. Pressure waves from sound travelling up the scala vestibuli and back down the scala tympani produce a shearing force on the hair cells of the organ of Corti in the cochlear duct. The hairs (cilia) of the hair cells are imbedded in the gelatinous tectorial membrane (Fig. 2), which has a relatively high inertial resistance to movement, so that sound-induced shearing forces bend the hairs. This bending produces mechanical opening of ionic channels (Fettiplace, 1990), depolarizing the hair cells due to K⁺ influx. Within the cochlea, hair cell sensitivity to frequencies progresses from high frequencies at the base to low frequencies at the apex. The cells in the single row of inner hair cells passively respond to deflections of sound-induced pressure waves. Cells in the rows of outer hair cells can elongate or shorten in response to the motion of the basilar membrane to actively produce amplification or attenuation of the response of the inner hair cells (Møller, 1993). Efferent innervation by fibres from the olivary nucleus caudally and the dorsal nucleus of the trapezoid body rostrally also provide regulation of the sensitivity of the inner and outer hair cells (Liberman, 1991). The scala vestibuli and scala tympani are filled with perilymph, similar in composition to extracellular fluid, while the cochlear duct is filled with endolymph, similar in composition to intracellular fluid.

Fig. 1. Schematic diagram of a cross-section of the cochlea, demonstrating the scala vestibuli, scala tympani and scala media or cochlear duct. The organ of Corti rests on the basilar membrane, with the hair cell cilia imbedded in the gelatinous tectorial membrane. The outer margin of the cochlear duct contains the stria vascularis. (From Bloom, W. & Fawcett, D.W. (1975). A Textbook of Histology, 10th edn. Philadelphia: W.B. Saunders. Reproduced with permission.)
fluid. The hair cells synapse on processes of neurons of the spiral ganglia, initiating signal transmission into the central nervous system via the eighth cranial nerve. On the lateral wall of the cochlear duct is the stria vascularis, a three-cell-layer thick vascularized epithelium not bounded by a basal lamina (Santi, 1988). The tissue is rich in Na⁺-K⁺-ATPase, responsible for the secretion of high levels of K⁺ into the endolymph of the cochlear duct. Also present in the stria vascularis are melanocytes, which appear to be critical to the maintenance of the stria (see below) but whose function at this site is unknown.

**CLASSIFICATION OF DEAFNESS**

Peripheral deafness (or hearing loss), defined as that being due to abnormalities outside the central nervous system (CNS), can be characterized by three pairs of descriptors: inherited or acquired, congenital or later-onset, and sensorineural or conductive. This results in eight classifications of deafness, but only three are commonly seen in dogs and cats: inherited congenital sensorineural, acquired later-onset sensorineural and acquired later-onset conductive. Inherited congenital sensorineural deafness is usually, but not always, associated with pigmentation genes responsible for white in the coat. Acquired later-onset sensorineural deafness is most often associated with ototoxicity or ageing-related hearing loss (presbycusis), but can also result from otitis interna, noise and other causes. Acquired later-onset conductive deafness is associated with chronic otitis externa and media or excess cerumen production. No forms of inherited late-onset deafness, either sensorineural or conductive, have been identified in dogs or cats, but the conditions are seen in humans. Acquired congenital deafness, either sensorineural or conductive, may result from malformations, intrauterine infections or drug toxicity, or anoxia, but these are not common. Causes of acquired deafness are listed in Table I.

Central deafness can theoretically result from a variety of retrocochlear lesions, but in practice is rare. The auditory pathways co-mingle information from both ears from the level of the cochlear nuclei rostrally, so it is difficult to produce total

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Fig. 2. Drawing of the organ of Corti, demonstrating the inner and outer hair cells and the spiral ganglion cells that become the cochlear nerve. (From Pansky, B. & Allen, D.J. (1980). *Review of Neuroscience*. New York: Macmillan. Reproduced with permission.)
unilateral central deafness, and bilateral central deafness requires lesion of a significant portion of the brain-stem or mid-brain, or a bilateral lesion of auditory cortex. Significant signs beyond deafness would accompany lesions of this sort.

Deafness can also be classified as either syndromic or non-syndromic. It has not been established whether deafness in dogs and cats associated with white hair coat colour and blue irises can be considered a form of syndromic deafness. The deafness seen in Dalmatians and white cats is often likened to Waardenburg syndrome (Foy et al., 1990; Baldwin et al., 1992), a dominantly-inherited condition in humans with incomplete penetrance consisting of deafness, blue irises, a stripe of white in the hair and beard with premature greying, and minor structural facial deformities.

**AETIOLOGIES OF DEAFNESS**

*Congenital sensorineural deafness*

The earliest studies of deafness in animals were in the Dalmatian in the last century (Rawitz, 1896); most studies have been performed with Dalmatians or white cats. Deafness does not develop in dogs and cats until the first few weeks of life, with normal functional development occurring to that point (Pujol & Hilding, 1973). Studies in our laboratory have shown that Dalmatians do not go deaf until weeks 3–4 after birth. The histological pattern that occurs in most dog breeds and white cats is known as cochleo-saccular, or Scheibe, type of end organ degeneration. The deafness results from initial degeneration of the stria vascularis, followed by the collapse of Reissner's membrane and the cochlear duct, degeneration of the hair cells of the organ of Corti and collapse of the saccule. Secondary loss of spiral ganglion cells is also seen at later stages (Lurie, 1948; Hudson & Ruben, 1962; Bosher & Hallpike, 1965; Anderson et al., 1968; Suga & Hattler, 1970; Igarashi et al., 1972; Johnsson et al., 1973; Mair, 1973, 1976). The cochlear hair cell and spiral ganglion cell loss is permanent in mammals. Histological

### Table I

<table>
<thead>
<tr>
<th>Cause</th>
<th>Time of onset</th>
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<tbody>
<tr>
<td></td>
<td>Congenital</td>
</tr>
<tr>
<td>Infections</td>
<td>X</td>
</tr>
<tr>
<td>Otitis (externa/media/interna)</td>
<td>X</td>
</tr>
<tr>
<td>Meningitis</td>
<td>X</td>
</tr>
<tr>
<td>Ototoxicity (drug, chemical)</td>
<td>X</td>
</tr>
<tr>
<td>Anoxia</td>
<td>X</td>
</tr>
<tr>
<td>Anaesthesia</td>
<td>X</td>
</tr>
<tr>
<td>Noise</td>
<td>X</td>
</tr>
<tr>
<td>Malformations (agenesis, ossicle fusion)</td>
<td>X</td>
</tr>
<tr>
<td>Trauma</td>
<td>X</td>
</tr>
<tr>
<td>Presbycusis</td>
<td>X</td>
</tr>
<tr>
<td>Unknown</td>
<td>X</td>
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</table>
studies of deaf Dalmatians have shown that the degeneration begins as early as 1 day after birth, and is clearly evident histologically by 4 weeks (Johnsson et al., 1973). Degeneration begins in the middle coil of the cochlea, followed by the basal then apical coils (Anderson et al., 1968). The cause of the strial degeneration is not known, but there is an observed absence of melanocytes in the strial tissue of many deaf animals (Savin, 1965; Steel et al., 1987). The function of melanocytes in normal stria is not known but appears to tie in with hair pigment associations with deafness (Steel & Barkway, 1989; Carlisle et al., 1990). Most melanocytes originate in the neural crest (Weston, 1969), so the absence of strial melanocytes could reflect either a failure of migration from the neural crest or a failure of differentiation after arrival. In the Doberman, and probably other dog breeds not carrying the merle or piebald pigment genes, the deafness results from direct loss of cochlear hair cells without any antecedent effects on the stria vascularis (Wilkes & Palmer, 1992).

It has been shown that the auditory cortex of deaf Dalmatians is grossly reduced in size (Ferrara & Halnan, 1983), leading the authors to the suggestion that the origin of deafness in the breed was central rather than peripheral. Although not reported, it is likely that other CNS structures in the auditory pathway were also smaller than in hearing animals. However, it is well known from classical studies that kittens whose eyelids were kept sealed after birth failed to develop normal CNS visual structures, demonstrating that normal sensory input is necessary for the full development and maintenance of these structures (Hubel et al., 1977). As a result, the findings in the Dalmatian are undoubtedly a reflection of a similar pathophysiological process. These CNS changes in deaf dogs have been used to justify euthanasia on the basis of having an 'abnormal' brain, but neurologically the brain function of deaf animals is normal except for the loss of auditory function.

**Conductive deafness**

Conductive deafness may result from developmental defects affecting the ossicles (such as fusion), from failure of the ear canal to completely open after birth or from otosclerosis, but these events have not been documented in dogs or cats and are probably rare. Congenital tympanic membrane absence occasionally occurs, but does not produce deafness. Conductive deafness is most often a result of chronic otitis externa and media, where stenosis and eventual occlusion of the external canal results, or impaction from excess cerumen accumulation. Chronic otitis externa may ultimately result in mineralization and ossification of the external ear canal, requiring lateral ear resection (Elkins et al., 1981) or other remedies. However, hearing function can be maintained, or even regained, after procedures as extreme as total ear canal ablation with lateral bulla osteotomy (Payne et al., 1989; Krahwinkel et al., 1993).

**Ototoxicity**

Ototoxic agents may cause hearing loss or deafness by direct effects on cochlear and/or vestibular hair cells, or may cause damage to the stria vascularis with secondary hair cell loss (Miller, 1985). Ototoxicity in humans is frequently accompanied by tinnitus, a high pitched ringing in the ears. Ototoxicity in dogs
and cats may likewise be accompanied by behaviour suggesting the presence of similar sensory phenomena. Over 180 compounds and classes of compounds have been identified as ototoxic (Govaerts et al., 1990; Mansfield, 1990; Pickrell et al., 1993). Many of those most likely to be seen in veterinary practice are listed in Table II; it must be noted that not all are equally toxic. In some cases the ototoxic effects are reversible if caught early, such as with salicylates, but in most instances the deficit is permanent by the time of detection. The best recognized, and perhaps most frequent, agents of ototoxicity are the aminoglycoside antibiotics, especially gentamicin (Govaerts et al., 1990). Drugs within this group are also nephrotoxic, and vary in their toxicity to the auditory, vestibular and renal systems. Gentamicin and streptomycin are most toxic to the vestibular system, while neomycin, kanamycin, tobramycin and amikacin are most toxic to the cochlea; nitrofurazone is thought to be the least toxic (Govaerts et al., 1990). However, since gentamicin and neomycin are the most frequently used aminoglycosides in veterinary practice, especially as topical otic agents, they are the drugs most likely to produce cochlear ototoxicity in companion animals.

The mechanism of toxicity of aminoglycosides is unclear, but the pathology includes a progression from basal coil outer hair cells, to more apical outer hair cells, followed by inner hair cells; strial changes are concurrent with, or precede outer hair cell changes (Govaerts et al., 1990). Although the effect is often ascribed to concentration of the drug in the perilymph, the most current evidence points to binding of the drug to glycosaminoglycans of the stria vascularis and disruption of phosphoinositide metabolism (Govaerts et al., 1990). It has been reported that serum gentamicin levels must exceed a 2 μg ml⁻¹ threshold level for

<table>
<thead>
<tr>
<th>Table II</th>
<th>Selected ototoxic drugs and chemicals</th>
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<tbody>
<tr>
<td><strong>Aminoglycoside antibiotics</strong></td>
<td><strong>Non-aminoglycoside antibiotics</strong></td>
</tr>
<tr>
<td>Amikacin</td>
<td>Amphotericin B</td>
</tr>
<tr>
<td>Dibekacin</td>
<td>Ampicillin</td>
</tr>
<tr>
<td>Framycetin</td>
<td>Bacitracin</td>
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<tr>
<td>Gentamicin</td>
<td>Chloramphenicol</td>
</tr>
<tr>
<td>Kanamycin</td>
<td>Chlorotetracycline</td>
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<tr>
<td>Neomycin</td>
<td>Colistin</td>
</tr>
<tr>
<td>Netilmicin</td>
<td>Erythromycin</td>
</tr>
<tr>
<td>Sisomicin</td>
<td>Griseofulvin</td>
</tr>
<tr>
<td>Streptomycin</td>
<td>Hygromycin B</td>
</tr>
<tr>
<td>Tobramycin</td>
<td>Minocycline</td>
</tr>
<tr>
<td>Diuretics</td>
<td>Polymixin B</td>
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<tr>
<td>Bumetanide</td>
<td>Tetracyclines</td>
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<tr>
<td>Furosemide</td>
<td>Vancomycin</td>
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<tr>
<td>Ethacrynic acid</td>
<td>Antiseptics</td>
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<tr>
<td>Antineoplastic agents</td>
<td>Benzalkonium chloride</td>
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<tr>
<td>Actinomycin C&amp;D</td>
<td>Benzethonium chloride</td>
</tr>
<tr>
<td>Cisplatin</td>
<td>Centrime</td>
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<tr>
<td>Nitrogen mustard</td>
<td>Chlorhexidine</td>
</tr>
<tr>
<td>Vinblastine</td>
<td>Ethanol</td>
</tr>
<tr>
<td>Vincristine</td>
<td>Iodine &amp; iodophors</td>
</tr>
</tbody>
</table>

Data from Govaerts et al. (1990), Mansfield (1990) and Pickrell et al. (1993).
over 10 days to produce toxicity (Sande & Mandell, 1990). Early ototoxic effects of gentamicin may be reversible by calcium administration (Pickrell et al., 1993).

Route of administration may affect ototoxicity, with systemic exposure providing better access of drugs to the cochlea than topical administration in ears with intact tympanic membranes. However, tympanum rupture frequently accompanies otitis externa, increasing access of drugs to the oval and round windows of the cochlea, through which absorption occurs. As a result, care must be exercised in topical drug application when visualization of the tympanic membrane is not possible, and when possible it is advisable to monitor hearing function with brain-stem auditory evoked response (BAER) recordings when high concentrations and long treatment courses of gentamicin or similar agents are employed. Attempts by us to produce ototoxicity in dogs with both intact and ruptured tympanic membranes, using typical clinical treatment protocols and assessing toxicity with BAER recordings, were unsuccessful for both gentamicin (Strain et al., 1995) and chlorhexidine (Merchant et al., 1993). This suggests, but does not guarantee, that topical application of drugs at recommended levels can generally be assumed to be safe. Age, concurrent infection, anaesthesia, or preexisting cochlear damage may potentiate drug ototoxicity, and repeated courses of antibiotic treatment may produce cumulative effects that are initially clinically not apparent.

Presbycusis

Presbycusis is the decline in hearing associated with various types of auditory system dysfunction that accompany ageing, and cannot be accounted for by otorotraumatic, genetic or pathological conditions (Schuknecht, 1955; Willott, 1991). Presbycusis can be classified into four types of pathology: sensory, neural, strial and cochlear conductive (Schuknecht & Gacek, 1993). The pathological change in most dogs and cats appears to be sensorineural, although decreased tympanum and ossicle joint articulation flexibility can potentially contribute. Presbycusis is common in geriatric dogs (Knowles et al., 1988, 1989), but prevalence rates or other related data are not available. Although it is a progressive disorder, owners usually report an acute onset because of the ability of the animal to compensate for hearing loss until nearly complete deafness occurs. Hearing aids have been successfully utilized in dogs with some residual auditory function, but not all dogs will tolerate the presence of the ear plug (Marshall, 1990). The primary determinant of the success of hearing aids is the ability of the owner to train the animal to accept the presence of a foreign body in the ear canal. Because of this training requirement and the sensitivity of the cat to ear contact it is unlikely that these devices would be successful in cats. There is no known way of retarding the progression of the deafness. In humans, men are affected more at high frequencies, while women are affected more at low frequencies (Jerger et al., 1993), but it is not known if similar patterns hold in dogs or cats.

Noise

Noise-induced hearing loss or deafness can be temporary or permanent (Peterson, 1980). Temporary increases in hearing threshold occur after brief exposure to intense sounds (over 100 dB), with gradual recovery of function
occurring over periods ranging from minutes to 2 weeks. Noise-induced hearing loss is thought to result from either disarrangement or breakage of hair cell cilia (Flottorp, 1990), but can also result from damage to the tympanum and ossicles. Continuous or repeated exposure to noise results in a progressive loss of hair cells and a corresponding deafness. In humans, the greatest hearing loss is at the middle frequency range near 4000 Hz, but progresses to higher and lower frequencies with continued exposure (Peterson, 1980). Dogs used to hunt with firearms, like their human companions, may develop noise-induced hearing loss (personal observation).

Other

Hearing loss or deafness may also result from anoxia, anaesthesia, trauma or infections, such as otitis interna and meningitis. On occasion, interactive effects may be expected to produce loss or deafness when the individual causes would have been insufficient alone to produce an effect (Pickrell et al., 1993).

PREVALENCE

Dogs

Congenital deafness in the dog has been reported in the literature (Erickson et al., 1977; Hoerlein, 1978; Clark & Stainer, 1983; de Lahunta, 1983; Oliver et al., 1987; Neer, 1990; Braund, 1994) and observed by the author in at least 54 breeds (Table III). Because of the various possible acquired causes of congenital deafness, and in the absence of breeding studies, it cannot be stated that all or even

<table>
<thead>
<tr>
<th>Dog breeds reported with congenital deafness</th>
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<tbody>
<tr>
<td>Akita</td>
</tr>
<tr>
<td>American Staffordshire Terrier</td>
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<tr>
<td>Australian Cattle Dog</td>
</tr>
<tr>
<td>Australian Shepherd</td>
</tr>
<tr>
<td>Beagle</td>
</tr>
<tr>
<td>Bichon Frist</td>
</tr>
<tr>
<td>Border Collie</td>
</tr>
<tr>
<td>Boston Terrier</td>
</tr>
<tr>
<td>Boxer</td>
</tr>
<tr>
<td>Bulldog</td>
</tr>
<tr>
<td>Bull Terrier</td>
</tr>
<tr>
<td>Catahoula Leopard Dog</td>
</tr>
<tr>
<td>Chow Chow</td>
</tr>
<tr>
<td>Cocker Spaniel</td>
</tr>
<tr>
<td>Collie</td>
</tr>
<tr>
<td>Dalmatian</td>
</tr>
<tr>
<td>Dappled Dachshund</td>
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<tr>
<td>Doberman Pinscher</td>
</tr>
</tbody>
</table>

most of these are inherited. Those breeds with the highest prevalence include Australian Cattle Dog, Australian Shepherd, Bull Terrier, Catahoula, Dalmatian, English Cocker Spaniel, English Setter and West Highland White Terrier.

Studies of the prevalence of deafness in dogs are limited; published findings have ranged from 0.065% (260 cases out of 397,235 canine hospital visits, United States, Mulvihill & Hanson, 1979) to 0.025% (272 cases out of 1.1 million canine hospital visits, United States, Hayes et al., 1981), to 0.875% (12 cases out of 1371 dogs reported in a survey of abnormalities in Australian purebreed dogs, Johnston & Cox, 1970). However, these numbers reflect only bilateral deafness, since they predate the widespread availability of electrodiagnostic hearing testing instrumentation that enables detection of unilateral deafness (see below). These numbers are probably low by at least a factor of four. Hearing testing has been adopted and promoted by several dog breed organizations, most notably the Dalmatian Club of America, the Bull Terrier Club of America and the English Setter Association of America. Similar European breed organizations have begun similar efforts. The prevalence in highly at-risk breeds for which data have been collected by the author and collaborators is shown in Table IV. These reported prevalence numbers may be low, since much of the data was collected at breed speciality dog shows, where deaf dogs and dogs not of show quality would have been excluded. Prevalence is highest in the Dalmatian, where 8.0% are bilaterally deaf and 21.8% are unilaterally deaf (Holliday et al., 1992; Strain et al., 1992).

Table IV

<table>
<thead>
<tr>
<th>Breed</th>
<th>Dogs tested</th>
<th>Bilaterally deaf</th>
<th>Unilaterally deaf</th>
<th>Bilaterally deaf</th>
<th>Total deaf</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dalmatian</td>
<td>4566</td>
<td>70.2% (3206)</td>
<td>21.8% (993)</td>
<td>8.0% (367)</td>
<td>29.8% (1360)</td>
</tr>
<tr>
<td>Bull Terrier</td>
<td>507</td>
<td>89.0% (451)</td>
<td>10.3% (52)</td>
<td>0.8% (4)</td>
<td>11.0% (56)</td>
</tr>
<tr>
<td>White</td>
<td>269</td>
<td>81.0% (218)</td>
<td>17.5% (47)</td>
<td>1.5% (4)</td>
<td>19.0% (51)</td>
</tr>
<tr>
<td>Coloured</td>
<td>237</td>
<td>97.9% (232)</td>
<td>2.1% (5)</td>
<td>0.0% (0)</td>
<td>2.1% (5)</td>
</tr>
<tr>
<td>English Setter</td>
<td>370</td>
<td>84.9% (314)</td>
<td>12.7% (47)</td>
<td>2.4% (9)</td>
<td>15.1% (56)</td>
</tr>
<tr>
<td>English Cocker Spaniel</td>
<td>388</td>
<td>91.2% (354)</td>
<td>7.0% (27)</td>
<td>1.8% (7)</td>
<td>8.8% (34)</td>
</tr>
<tr>
<td>Australian Cattle Dog</td>
<td>70</td>
<td>88.6% (62)</td>
<td>8.5% (6)</td>
<td>2.9% (2)</td>
<td>11.4% (8)</td>
</tr>
</tbody>
</table>

Table V

<table>
<thead>
<tr>
<th>Cat breeds carrying the white (W) coat pigment gene (Gebhard et al., 1979) and at risk for congenital deafness</th>
</tr>
</thead>
<tbody>
<tr>
<td>White</td>
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</table>

Prevalence in the Bull Terrier, English Setter, English Cocker Spaniel and Australian Cattle Dog is one-half to one-third that of the Dalmatian. Unilateral or bilateral deafness has been reported to occur in 75% of all white Norwegian Dunkerhounds, but the prevalence in coloured dogs is unknown (Foss, 1981). In Dappled Dachshunds, 54.6% are reported to be deaf, with 18.2% bilaterally deaf and 36.4% unilaterally deaf (Reetz et al., 1977).

Cats
Few cat breeds are noted for congenital deafness. Those reported with congenital deafness, or with potential for it, include all those carrying the dominant white (W) gene (Gebhardt et al., 1979; Table V) and perhaps the white spotting or piebald (S) gene (see below). Although cat owners in these breeds are familiar with the problem of deafness, little specific published information is available by breed. Several studies have examined deafness in mixed-breed white cats (Bosher & Hallpike, 1965; Bergsma & Brown, 1971; Mair, 1973; reviewed by Delack, 1984). Out of 256 white cats from these three studies, 12.1% were unilaterally deaf and 37.9% were bilaterally deaf, that is, a total of 50% were affected (Delack, 1984). When cats that were the offspring of two white parents were examined, the prevalence of deafness (unilateral or bilateral) ranged from 52-96%. When Mair (1973) and Bergsma & Brown (1971) examined the effect of blue eye colour on deafness, they found, respectively, a prevalence of deafness (unilateral and bilateral combined) of 85% and 64.9% in cats with two blue eyes, 40% and 39.1% in cats with one blue eye, and 16.7% and 22% in cats with no blue eyes. The author is unaware of any study of deafness in cats by specific breed. Pure-bred white cats are said to have a lower prevalence of deafness than mixed-breed white cats (Pedersen, 1991), but supporting data are unavailable.

GENETICS
It is usually impossible to determine the cause of congenital deafness unless a clear problem has been observed in a breed or carefully planned breedings are performed. In affected breeds, deafness has often been long-established but kept hidden from outsiders to protect reputations. Hereditary deafness can potentially result from any of several mechanisms: autosomal dominant or recessive, X-linked, mitochondrial or polygenic; in most instances the mechanism is unknown. Incomplete penetrance, where not all aspects of a deafness syndrome are expressed in an affected individual, frequently complicates an understanding of the mode of inheritance. No known X-linked or mitochondrial deafness has been reported in dogs or cats. With a few known exceptions, hereditary deafness is usually associated with pigmentation patterns, where increasing amounts of white in the hair coat increase the likelihood of deafness.

Dogs
Two pigmentation genes are often associated with deafness in dogs: the merle gene (seen in the Collie, Shetland Sheepdog, Dappled Dachshund, Harlequin Great Dane, American Foxhound, Old English Sheepdog and Norwegian...
Dunkerhound among others) and the piebald or extreme piebald gene (Bull Terrier, Samoyed, Greyhound, Great Pyrenees, Sealyham Terrier, Beagle, Bulldog, Dalmatian and English Setter). Not all breeds with these genes have been reported to be affected with deafness.

The merle (dapple) gene (M) produces a mingled or patchwork combination of dark and light areas (Little, 1957; Searle, 1968). This gene is dominant so that heterozygous dogs (Mm) show the pattern, which is considered desirable in many breeds. However, when two dogs with merle are bred, 25% on average will end up with the MM genotype. These dogs usually have a solid white coat and blue irises, are often deaf and/or blind and are sterile. Experienced breeders of these dogs know not to breed merle to merle. Heterozygous merles can also be deaf, with the likelihood of deafness increasing with increasing amounts of white in the hair coat. In this case the deafness is neither dominant nor recessive, but is linked to a dominant gene that disrupts pigmentation and secondarily produces deaf dogs.

Genetic transmission of deafness in dogs with the piebald (s') and extreme piebald (s") pigment genes, such as the Dalmatian, is less clear. These genes effect the amount and distribution of white areas on the body (Little, 1957; Searle, 1968). The canine piebald genes are recessive, but individuals in breeds such as the Dalmatian are homozygous, so all dogs within the breed express the pigment pattern. Deafness in Dalmatians does not appear to be dominant since deaf puppies result from hearing parents. It does not appear to be a simple recessive disorder: we have repeatedly bred pairs of deaf Dalmatians from our research colony and obtained many bilaterally hearing puppies, when all should have been deaf if the disorder was recessive. These findings might be explained by a polygenic cause, the presence of two different autosomal recessive deafness genes, or a syndrome with incomplete penetrance. Suggestions have been made for two different recessive genes, either of which can cause deafness, or two recessive genes where both are required to cause deafness (Hewson-Fruend, 1990), or a recessive multifactorial gene with incomplete penetrance (Greibrokk, 1994). Deafness is still clearly linked to the extreme piebald gene in Dalmatians. In this breed, the underlying coat colour is black (B) or liver (b, simple recessive). The extreme piebald gene (s") covers the colour with white, and the dominant ticking gene (T) opens the spots through the white. In Dalmatians with a patch, the s" gene does not completely suppress the underlying coat colour; the s" gene is only weakly expressed. Patched Dalmatians have been shown to have significantly lower deafness rates (Strain et al., 1992), but a patch is not allowed in the breed standard. Conversely, blue-eyed Dalmatians, where the normal brown iris pigment is suppressed, are significantly more likely to be deaf (Strain et al., 1992; Greibrokk, 1994). Blue eyes are allowed in the breed standard of the United States, but not in Canada or Europe. Dalmatians that are the offspring of one bilaterally hearing parent and one unilaterally deaf parent are twice as likely to be deaf (unilaterally or bilaterally) as dogs that are the offspring of two bilaterally hearing parents (Strain, 1992b). Efforts through breedings to reduce blue eyes in Norwegian Dalmatians reduced the prevalence of deafness (Greibrokk, 1994).

Recent studies have shown that deafness in Dobermans, which do not carry the merle or piebald genes, results from direct loss of cochlear hair cells without any effects on the stria vascularis (Wilkes & Palmer, 1992). Vestibular system signs,
including head tilt and circling, are seen, and the deafness is transmitted by a simple autosomal recessive mechanism. A similar pathology has been described for the Shropshire Terrier (Igarashi et al., 1972).

Numerous references report that most congenital deafness in dogs is autosomal recessive. However, the available data suggest that this is not true for most breeds.

**Cats**

The white (W) pigment gene in cats is autosomal dominant over colour, and is unrelated to albinism (Little, 1957; Searle, 1968). Cats carrying the W gene are not always solid white, often having coloured spots on their heads that may disappear with age. Unlike dogs with the merle gene, homozygous white cats do not have visual or reproductive defects, but they are more prone to the occurrence of blue irises and deafness, either unilateral or bilateral, and deafness occurrence increases with the number of blue eyes (Delack, 1984). Whether the cat is heterozygous or homozygous for W, the blue eyes and deafness have incomplete penetrance. Long-haired cats have a higher prevalence of blue eyes and deafness than short-haired cats (Mair, 1973). White cats carrying the underlying c Siamese dilution pigment gene can have blue eyes without deafness, and it has been suggested that the presence of this gene explains why pure-breed white cats are less often deaf than mixed-breed white cats (Pedersen, 1991). The white gene is present in many cat breeds (Table V), but no data are available on relative rates of occurrence of deafness between them.

A dominant piebald gene (S) is also found in various cat breeds (Pedersen, 1991; Searle, 1968), but there has been no report of deafness associated with its presence.

**DIAGNOSIS**

Since the ear canal does not open until approximately 5 days in cats and 14 days in dogs, and deaf puppies and kittens cue from the responses of littermates, it is not uncommon for deafness to go unrecognized for many weeks. In some breeds, bilaterally deaf puppies will display more aggressive play with littermates because they do not hear cries of pain, and both puppies and kittens after weaning will not waken at feeding time unless jostled. Bilateral deafness can usually be detected by behavioural testing with sound stimuli presented outside of the visual field or with the animal blindfolded, taking care to avoid visual or vibratory cues. The minimum desired response is a Preyer's reflex, or twitch of the ears in response to the sound; many animals will also orientate to the sound source. However, hearing animals, especially the young, quickly adapt and stop responding, resulting in equivocal results. Further, unilateral deafness cannot be detected by these measures; at best such animals may demonstrate difficulty in localizing the origin of a sound.

Objective assessment of the presence of auditory function requires a test known variously as the BAER, brain-stem auditory evoked potential (BAEP), or auditory brain-stem response (ABR). In this test, a computer-based system detects electrical activity in the cochlea and auditory pathways in the brain in much the same way
that an antenna detects radio or TV signals or an ECG detects electrical activity of the heart (Sims & Moore, 1984a; Sims, 1988; Strain, 1992a). The response waveform consists of a series of peaks identified by Roman numerals: peak I is produced by the cochlea and cochlear nerve, and later peaks are produced within the brain (Fig. 3(a)). The response is collected with a special computer through small subdermal electrodes. The most common electrode montage consists of one in front of each ear, one at the top of the head and one between and just caudal to the eyes. It is uncommon for an animal to show any evidence of pain from the placement of the electrodes, but it may object to the restraint and the irritation of wires hanging in front of its face. Recordings from cats often require a cat bag or other form of restraint. The stimulus click produced by the computer is directed into the ear with a foam insert earphone or headphones. Because of the microvolt amplitude of the response, a computer must average responses to a large number of stimuli (typically 1000) to unmask them from the scalp electroencephalogram (EEG) and electromyogram (EMG) activity within which the waveform is buried. Each ear is tested individually, and the test is usually complete in 10–15 min. When dogs or white cats are screened for congenital inherited deafness, a single sound intensity is generally used by examiners since the deafness, when present in an ear, is total; 95 dB nHL (normal hearing level) is used by the author, but no consensus standard has been developed. Assessment of partial hearing loss is usually evaluated at several intensities to gauge the extent of loss. Sedation or anaesthesia are generally unnecessary unless the animal becomes extremely agitated, which can usually be avoided with patient and gentle handling. However, the response is unaffected by chemical restraint, so some practitioners routinely employ sedatives. With complete peripheral deafness, peak I of the BAER is totally absent, as are the subsequent peaks (Fig. 3(b)). With partial hearing loss, as is seen with presbycusis and some cases of ototoxicity, the time to occurrence of peak I is increased and the amplitude of the peaks is diminished (Fig. 3(c)). The BAER changes during post-natal development of the auditory system, so appropriate reference values must be used when evaluating young animals (Buchwald & Shipley, 1986; Strain et al., 1991). BAER testing of puppies is not usually performed before 5 weeks of age, when the strial and hair cell degeneration of congenital deafness is complete. Many breeders of at-risk dog breeds routinely test entire litters before placing the dogs, and bilaterally deaf puppies are frequently euthanatized.

Animals that test as deaf with the BAER, but in whom conductive deafness is suspected, can be further evaluated by BAER using a bone stimulator instead of air-conducted clicks (Strain et al., 1993). A vibratory stimulus transducer is firmly held against the skull, preferably over the mastoid process, and BAER recordings are obtained in the usual manner. The auditory stimulus travels through bone to the cochlea, bypassing the outer and middle ears. When the deafness is conductive, a normal-appearing response is recorded (Fig. 3(d)). Because there are no known forms of inherited conductive deafness in dogs or cats, animals with this form of deafness are not precluded from breeding considerations.

Other diagnostic tests of auditory function are also available, but may be more difficult to employ, require additional high-cost equipment, or require anaesthesia. Impedance audiometry (Penrod & Coulter, 1980; Sims, 1988) permits
Fig. 3. Brain-stem auditory evoked responses recorded from dogs. (a) Air-conducted response recorded from a normal adult. (b) Air-conducted response recorded from a deaf Dalmatian puppy. (c) Air-conducted response recorded from a 13-year-old Boston Terrier with presbycusis. Note the delayed peak latencies and decreased peak amplitudes compared to (a). (d) Bone-conducted response from a normal adult. Latencies are shorter than in (a) because bone conduction transmission time is shorter than the air transmission time in the insert earphone tubing.

assessment of middle ear function. The middle latency auditory evoked potential (Sims & Moore, 1984b), similar to the BAER, tests auditory pathways up through the auditory cortex. Most recently, it has been shown that the cochlea generates very low intensity otoacoustic emissions in response to auditory stimuli, a response thought to reflect the active processes of outer hair cells. Transiently evoked otoacoustic emissions can be used to assess cochlear function (Sims et al., 1994).

MANAGEMENT

Dogs and cats with unilateral deafness make excellent pets, with owners often unable to detect any impairment. However, owners of these animals should be discouraged from breeding them to prevent further affected animals and an ultimate increase in the prevalence of the disorder. Some animals will show directional localization deficits and may not awaken to sounds if sleeping with the good ear against the ground. Animals with late-onset acquired deafness generally adapt well, but precautions must be observed to prevent vehicular injury or death and bite injuries to humans, especially children, when deaf dogs are startled. Animals bilaterally deaf from both congenital and acquired causes place greater reliance
on visual and vibratory sensory information to cope with the loss of auditory input. Dogs are easily trained to hand signals and other visual cues, such as flashing porch lights; some cats can similarly be trained. Obedience-training shock collars set to the lowest shock level can be used for recall of dogs.

Despite the worry of animal owners and those concerned with animal rights, the quality of life of deaf dogs and cats is not demonstrably diminished. Likewise, these animals do not have diminished mental capacities, any more than the average deaf or blind human has diminished mental capacity. The brain responds to the loss of a sensory modality by various forms of plasticity, whereby CNS structures that would have received input from that sensory modality constrict and adjacent structures expand to take advantage of the available space (Hata & Stryker, 1994).

A dilemma often occurs when bilaterally deaf puppies are identified in a litter. The official position of the Dalmatian Club of America is that such animals should be euthanatized, and individual owners of other breeds often concur, but this position is not universally accepted. The recommendation for euthanasia is more difficult to accept after placement of a deaf dog in a home and the ensuing development of emotional attachments. Some variation may result from differences in personalities between breeds, but dogs that are bilaterally deaf from birth may develop anxious or aggressive personalities from continuously being startled. They are prone to vehicular deaths, may scare-bite and require significantly greater effort to rear and protect. It is not uncommon for these animals to end up in animal shelters because of the inability of owners to cope with the deficit.

Genetic counselling for owners of at-risk breed dogs and cats will be difficult until the mechanisms of inheritance are identified or a DNA blood test is developed. In general, unilaterally deaf animals should not be bred, since they have the genetic defect and will pass it on to their offspring. Some breeders view hearing as just one of the spectrum of desirable or undesirable markers evaluated in breeding decisions, but the high prevalence of deafness in at-risk breeds suggests that a higher premium should be placed on hearing status for the overall benefit of the breed.

**FUTURE DIRECTIONS**

Waardenburg syndrome in humans is a pigment-related deafness syndrome consisting of hypopigmentation in the hair, blue eyes, deafness and minor facial structural abnormalities; two subtypes of Waardenburg syndrome are recognized. The defective gene for type 1 Waardenburg syndrome, known as PAX3, has recently been located on the long arm of chromosome 2 (Foy et al., 1990; Baldwin et al., 1992), providing the possibility for diagnoses through DNA blood testing. Unfortunately, blood from deaf Dalmatians did not test as abnormal in one assay (A. Milunksy, personal communication, 1994). This may be the result of a difference in chromosomes, since dogs have 39 pairs of chromosomes compared to the 23 pairs in humans. Efforts are currently underway to isolate the normal canine PAX3 gene, clone it, and develop DNA testing procedures to determine if the same gene is responsible for pigmentation-related deafness in dogs. More
recently, the defective gene for the human type 2 Waardenburg syndrome, named MITF, has been localized to the short arm of chromosome 3 (Tassabehji et al., 1994). DNA is being isolated from deaf dogs in our laboratory and stored for future work in this area.

In the absence of a reliable test to identify carriers of deafness genes in dogs, efforts are currently underway to establish a hearing registry in the United States, whereby dogs certified to have normal bilateral hearing would be registered to enable breeders to reliably select mates for their own animals to minimize the production of deaf offspring.

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