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References


Auditory Testing of Dogs with Inherited Retinal Degeneration

Gregory M. Acland,*† Roger R. Marsh,:‡ and Jerry W. Northington†

Auditory function was tested by brainstem-evoked response (BSR) audiometry in dogs affected by hereditary retinal degeneration (HRD). Comparison of BSR thresholds and latency–intensity functions revealed no significant difference between progressive rod–cone degeneration (PRCD) affected and unaffected miniature poodles, and no evidence of sensorineural hearing loss in HRD-affected English cocker spaniels and miniature schnauzers. The authors conclude that hearing loss is not a feature of the retinal degenerations in these dogs. Invest Ophthalmol Vis Sci 26:785–788, 1985

Hereditary retinal degenerations in the dog are known collectively as progressive retinal atrophy (PRA).¹ Specific forms of PRA have been well characterized in some breeds (eg, progressive rod cone degeneration (PRCD) in the miniature poodle) and are recognized but not fully characterized in other breeds.² The inference that PRA in different breeds represents different diseases has been proven in some cases³ and is supported by the different ages of onset and rates of progression of the syndrome among the different breeds. PRA in the miniature poodle, the English cocker spaniel, and the miniature schnauzer is recessively inherited. In poodles and spaniels, PRA is late in onset and slowly progressive, but in schnauzers the onset is earlier and the progression more rapid. Affected dogs exhibit night blindness progressing to total blindness, abnormal electroretinograms, morphologic abnormalities of the photoreceptors and, ophthalmoscopically, progressive retinal thinning, attenuation of the retinal vasculature and pallor of the optic disc.¹ In all these respects PRA mimics retinitis pigmentosa (RP), a syndrome of hereditary retinal degeneration of man.³

Hearing loss is the most frequently noted extracocular symptom of persons affected with RP.³ The association of sensorineural hearing impairment and RP defines Usher syndrome,⁴ although this diagnosis is often reserved for cases where the hearing loss is both congenital and profound.⁴ Nonetheless, regardless of the degree of hearing loss and the probability that some cases represent chance association, segregation analysis indicates that hearing impairment is significantly associated with RP is recessively inherited.⁵ In differing clinical populations, the incidence of hearing impairment among RP patients ranges from 20 to 30%.

Since PRA in the dog models RP in humans and because of the association between RP and hearing loss, we evaluated auditory function, by brainstem-evoked response audiometry (BSR), in dogs affected with PRA.

Materials and Methods. Animals: Seventeen miniature poodles, seven English cocker spaniels, and two miniature schnauzers were tested (Table 1). All were cared for, in a research colony for the study of
Table 1. Summary of dogs tested and results of BSR audiometry

<table>
<thead>
<tr>
<th>Breed</th>
<th>Disease (PRA status)</th>
<th>No. dogs tested</th>
<th>Age Mean</th>
<th>Age Range</th>
<th>BSR threshold* Mean</th>
<th>BSR threshold* SEM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Poodle</td>
<td>Affected</td>
<td>10</td>
<td>3.5 yr</td>
<td>8 mo–7 yr</td>
<td>6.5</td>
<td>2.4</td>
</tr>
<tr>
<td>Spaniel</td>
<td>Nonaffected</td>
<td>7</td>
<td>3.3 yr</td>
<td>11 mo–6 yr</td>
<td>7.2</td>
<td>2.3</td>
</tr>
<tr>
<td>Schnauzer</td>
<td>Affected</td>
<td>7</td>
<td>2.2 yr</td>
<td>1.0–6 yr</td>
<td>1.5</td>
<td>6.0</td>
</tr>
</tbody>
</table>

* Threshold is in dB relative to “normal” canine BSR threshold.

SEM: standard error of the mean.

dogs with HRD at the University of Pennsylvania, in adherence to the ARVO Resolution on the Use of Animals in Research. Ten poodles were affected with PRCD, and seven were homozygous normal or heterozygous. PRCD, in the colony from which these dogs arose, has been well described. All spaniels and both schnauzers were affected with PRA.

The retinal disease status of each dog was either assessed by morphologic examination of the retina from an enucleated eye or known by pedigree analysis (where the status of both parents was known and the mating could produce only one result).

**Testing procedure:** Prior to BSR audiometry each dog was sedated by intravenous injection of atropine (0.02 mg/kg), sparine (1.0 mg/kg), and morphine (2.0 mg/kg). Ears were cleaned and examined to be certain that the tympanic membrane was normal.

The auditory stimulus was a click generated by a 0.1 ms square pulse driving a small earphone inserted in the animal’s ear canal. This stimulus was presented 500 times at 10 times per second for each intensity tested. The click was first presented at an intensity known from clinical experience (JWN) to be 50 dB above the average BSR threshold of normal dogs. This intensity was then progressively reduced in 10–20 dB steps until no BSR response was detectable.

The BSR was recorded from subcutaneous needle electrodes at the vertex of the skull and over the mastoid of the stimulated ear. Each response signal was bandpass filtered (20 Hz–20 kHz), and 500 such responses were averaged by a Disa 14G11 electromyograph (Disa Electronics; Franklin Lake, NJ).

Although BSR audiometry is objective in that the subject need not judge if a stimulus is present, some judgment may be required by the examiner to identify a near threshold response in the presence of electrical interference. Thus BSR testing and analysis were performed with the examiner having no contact with the animals before they were sedated, and not knowing which animals were affected.

Two criteria of cochlear function were assessed: the BSR threshold (ie, the stimulus required to elicit a just detectable response) and the shape of the BSR latency–intensity function (see below).

**Results.** Identifiable responses were obtained from all dogs although the waveform varied among dogs. There was no discernible difference between the BSRs of normal and affected poodles (Fig. 1). The difference in mean threshold between affected and controls...
Fig. 3. Latency-intensity functions for PRA-affected spaniels (solid) and schnauzers (broken lines). There is variation between breeds, but there is no evidence of cochlear impairment.

(Table 1) was less than 1 dB and nonsignificant (t = 0.2106, p > 0.1, 15 degrees of freedom). BSR latency-intensity functions for these two groups were also convincingly similar (Fig. 2). The BSR threshold for spaniels and schnauzers was lower, with one exception, than the means for both PRCD-affected and unaffected poodles (ie, a response was elicited a 0 dB) (Table 1) and the latency intensity function (Fig. 3) was normal. The exception, a 6-year-old spaniel, had a threshold 35 dB above that of the mean for the other spaniels and at 50 dB her response latency (3.85 msec) was markedly prolonged.

Discussion. Assessment of hearing in dogs has been hindered by lack of a suitable objective test and by the inadequacies of subjective tests. BSR audiometry, a test of neuronal activity in the auditory pathways from the cochlea through the brainstem, does much to resolve this problem. The BSR is largely unaffected by the subject’s state of arousal, or sedative and anesthetic agents. If hearing is normal, stimuli less than 10 dB above absolute auditory threshold will elicit a measurable response. If hearing is abnormal, BSR audiometry can estimate the degree of loss. Conductive impairment (eg, middle ear dysfunction) may be distinguished from sensorineural hearing loss by analysis of the relationship between the stimulus intensity and the BSR response latency. A vertex-positive wave, attributable to the inferior colliculus, can be identified, even in responses to barely audible stimuli, in the BSR of various animals and is termed wave V in the BSR of humans. The manner of increase in latency of this wave with decreasing stimulus intensity (the BSR latency-intensity function) allows discrimination between conductive and sensorineural hearing impairment. In the former, latency increases gradually with decreasing intensity just as in the normal ear, but the latency-response function is shifted as increased sound levels are required to overcome the conductive impairment. Sensorineural hearing loss, however, is characterized by recruitment—the rapid growth in response as sound intensities rise above threshold. If, say, threshold were elevated by 50 dB, then a 45-dB sound would be inaudible, but a 65-dB sound might produce a nearly normal response. In the BSR, the latency of the response to loud sounds may be nearly normal, but the latency suddenly increases as intensity is reduced towards threshold.

BSR variation from dog to dog and between breeds reflects differences in the summation of responses from several areas in the brainstem. The relative input from an area depends on the orientation of its projections to the recording electrodes and its distance from them. Age, sex- and breed-matched controls, as were available for the poodles in this study, can eliminate much of this variability. Assessment of responses from spaniels and schnauzers was more subjective than for poodles, since such control dogs were unavailable. The criteria applied for normal sensorineural function in the spaniels and schnauzers were a smooth and gradual slope to the latency-intensity function (Fig. 3) and thresholds comparable to the poodles (Table 1). Only one dog was abnormal by these criteria and her impairment had the characteristics of conductive loss.

We conclude that hearing loss is not associated with PRCD in miniature poodles nor with the hereditary retinal degenerations present in this colony of English cocker spaniels and miniature schnauzers.

Key words: retinal degeneration, progressive rod-cone degeneration, retinitis pigmentosa, brainstem-evoked response audiometry, hearing, dogs

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