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Bilateral Later-Onset Sensorineural Deafness Diagnosed by Brainstem Auditory Evoked Response in a Border Collie

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Abstract : A 4-year-old, intact female Border Collie was presented for evaluation of hearing impairment. Clinical, neurological, otoscopic and magnetic resonance imaging examinations were carried out to determine the cause of hearing loss, but no remarkable change was found. Then, brainstem auditory-evoked response test was performed to assess hearing loss, and the dog had a bilateral sensorineural deafness was revealed. Since possible causes of acquired hearing loss were ruled out by several examinations and history taking, bilateral later-onset deafness was suspected to be genetic and not congenital. This report suggested the possibility that dogs had inherited later-onset sensorineural deafness.

Key words **:** Border Collie, brainstem auditory evoked response, canine, later-onset sensorineural deafness.

Introduction

The prevalence of hereditary deafness is substantial in many breeds of dogs such as Dalmatians, Border Collies, Australian Cattle dogs, Old English Sheep Dogs and Jack Russell Terriers (10,12,13). Some previous studies reported that 29.7% of 1031 Dalmatians and 2.8% of 2597 Border collies had congenital sensorineural deafness (CSD) (6,12). CSD results from loss of hearing receptors within a few weeks post-natally and is permanent in mammals (4,10).

Evaluation of hearing in dogs in the past was often conducted by measuring behavioral response to a sound or series of sounds, but it is often unreliable and subjective (2,13). To assess auditory function in dogs, electrodiagnostic testing should be employed as an alternative to behavioral testing (9). The brainstem auditory-evoked response (BAER) is one of the most frequently used testing modalities available because the test is objective, reasonably easy to perform, noninvasive, safe, and cost-effective (2,8,13).

In dogs, hereditary deafness has primarily been regarded as congenital and recessive. However, there is quite a lot of inherited middle-age hearing loss in humans and this late-onset hearing loss is more often inherited as a dominant condition (5,7). To our knowledge, there are no documented forms of hereditary deafness that are not congenital, except possibly for the Rhodesian Ridgeback and Border Collie (14). Here we described a case of bilateral later-onset deafness, which was suspected to be genetic. A Border Collie was found to be normal on clinical, neurological, otoscopic and magnetic resonance imaging (MRI) examinations, and was diagnosed with bilateral deafness following BAER testing.

Case

A 4-year-old, intact female Border Collie was presented to the Veterinary Medical Teaching Hospital of Seoul National University for evaluation of a bilateral hearing loss. The dog did not obey commands using sounds or wake from her deep

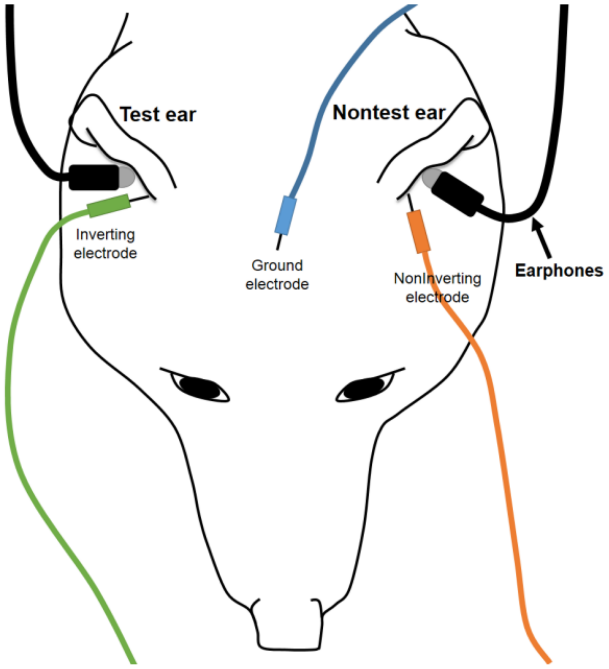

Fig 1. Schematic diagram of recording procedures for BAER.

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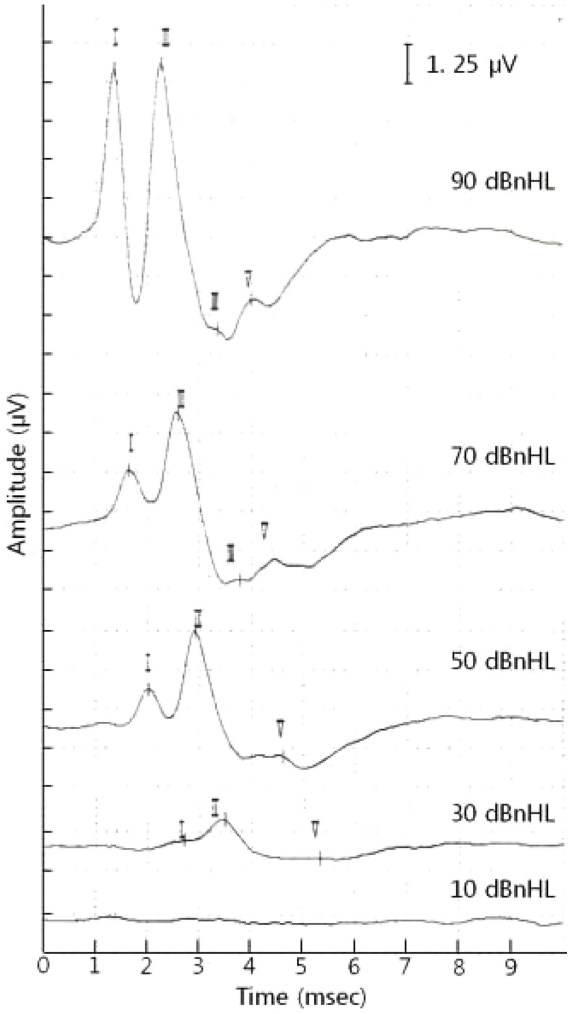

Fig 2. Change of BAER waveforms generated by monoaural stimulation in a control dog with normal hearing. Note that the amplitude of wave, especially wave I and II, is decreased depending on reduced simulation intensity. The BAER of this dog became isoelectric at the stimulation intensity of 10 dBnHL.

sleep despite noises for some time past. Its coat color was bicolor with black and white. The iris color of both eyes was brown.

The dog was found to be in good health on clinical examination. There was no evidence of ear pain or inflammation. Otoscopic examination revealed that both tympanic membranes were intact and ear canals were clean and grossly normal. MRI examination was performed to rule out the possibilities of brain lesions and other abnormalities, but no remarkable change was found on MRI. However, the dog responded ambiguously during behavioral assessment.

A complete BAER threshold estimation was performed to assess the dog's auditory function. Since BAER are not significantly affected by anesthetic drugs (2,8-10,13), the dog was under anesthesia for 10 min procedure with 6 mg/kg propofol (Provive 1%; Claris Lifesciences, Vasana, India), administrated intravenously. The dog's BAER was obtained using a

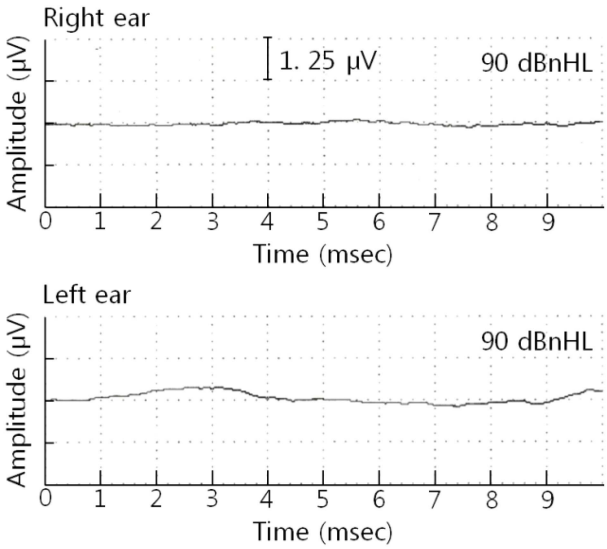

Fig 3. BAER recordings for right ear and left ear stimulations in a Border Collie. In this dog, the BAER waveform is flat at 90 dBnHL stimuli.

Neuropack 2 (Nihon Kohden, Tokyo, Japan) according to previously described methods (1,13). To detect each BAER, 3 platinum needle electrodes were placed subcutaneously. The inverting electrode was placed rostral to the targus of the test ear, the noninverting electrode was placed rostral to the targus of the nontest ear, and the ground electrode was placed on the vertex (Fig 1). To elicit each BAER, earphones were placed into both ears and the test ear was stimulated with alternating acoustic clicks while the non-test ear was masked with white noise (a broadband signal that contains all frequencies equally and prevents the nontest ear from contributing to the BAER). The clicks were driven by 0.1 msec square wave electrical impulses that produced broadband acoustic clicks containing frequencies between 50 Hz and 3000 Hz. The stimulating rate was 30/sec, averaging of 1000 sweeps was performed to record BAER. First, stimulus intensity was set at 90 dBnHL (decibels normalized hearing level) and the intensity level decreased in 5 dBnHL steps to determine the BAER threshold. The BAER threshold was defined as the lowest stimulus level at which a repeatable waveform could be obtained.

The BAER examinations of 8 mongrel dogs were also performed, and the BAER thresholds and BAER waveforms were recorded as controls. These dogs, 2 years old and clinically normal, had normal hearing and no neurological abnormalities from subjective observations. All animal procedures were performed in accordance with the guidelines of the Institutional Animal Care and Use Committee of Seoul National University (SNU-080916-2).

The BAER waveforms generated in response to the stimulation of both ears of control dogs were normal with decreased amplitude as stimulus intensity decreased (Fig 2). The BAER threshold average of these dogs was 31.25 ± 5.32 dBnHL. How-

ever, the response waveforms of a Border Collies appeared as flat lines without clearly defined peaks and valleys at 90 dBnHL (Fig 3). This absence of a response indicates that both ears being tested are non-functional and the dog is bilaterally deaf.

Discussion

Hearing impairment may be congenital or later-onset, and sensorineural or conductive (10). This case illustrates that a Border Collie might have later-onset sensorineural deafness. The major causes of hearing loss are aging, long-term exposure to environmental noise, genetic, disease, exposure to ototoxic chemicals and physical trauma (10). Geriatric hearing loss is estimated to begin at 8-10 years (14). However, adult-onset deafness in Border Collies often has an earlier onset (3-5 years) than deafness resulting from the physiological aging of hearing organs. The Border Collie in this case was 4 years old. Thus, presbycusis in this case should be excluded. Since the dog in this case appeared normal after clinical, neurological, otoscopic and MRI examinations, hearing loss by disease or illness can be also ruled out. The possibility of exposure to environmental noise or ototoxic chemical, or trauma was excluded through history taking. Taken together, hearing loss of the Border Collie in this case is more likely to deafness related to genetics. In dogs and cats, most inherited deafness is congenital, although there are human forms of inherited later-onset deafness (11). Inherited later-onset deafness is different from congenital deafness or geriatric hearing loss. Congenital deafness usually occurs 2-4 weeks after birth in dogs and cats (11). Because the owner of the dog in the present case used sounds such as whistles in training not so long ago and the dog had been performing training well, congenital deafness can be ruled out in this case. As a result, there's a strong possibility that hearing loss of this Border Collie may be inherited later-onset sensorineural deafness. This is the rare condition in dogs, and a recent previous study identified the discovery of a possible genetic link to later-onset hearing loss in Border Collie (14). In this study, candidate variants on CFA6 that are strongly associated with adult-onset deafness in Border Collie were identified.

In humans, later-onset hearing loss is more often inherited as a dominant condition. Since both parents of Border Collie in this case showed normal hearing, it would imply both parents were carriers of the mutated gene as in a recessive condition. However, a possible genetic link to later-onset deafness in Border Collie is yet lack. Some studies demonstrated that deaf Border Collies had higher observed rates of merle coat pigmentation and blue iris pigmentation than normal hearing Border Collies (3,6). Border Collies suffering from CSD carry the pigmentation allele associated with deafness including the allele S^{p} for piebald spotting, the allele S^{W} for extreme white piebald coloring, or the dominant merle gene M (6). However, the Border Collie in this case had black and white coat color with brown iris. Although a previous study observed robust associations, none of the candidate single-nucleotide polymorphisms tracked perfectly with adult-onset deafness in Border Collie (14). This is probably because adult-onset deafness in the Border Collie is multigenic trait. Since the mode of inheritance has not been established in Border Collies affected with later-onset deafness, future studies focusing on a few genes shown to cause later-onset deafness in humans and pedigrees of deaf Border Collies are needed.

It is important to realize the presence of inherited later-onset deafness in dogs and to understand the value of BAER testing in achieving a diagnosis of this condition. The possibility also exists that later-onset deafness may be present in other breeds, although none has been detected. One factor that may have made the disorder more notable in the Border Collie is that the dog's owner is attuned to changes in a dog's hearing since the dog depends so heavily on its hearing ability in training procedures. In this case, although behavioral assessment was not a reliable diagnostic test for the detection of bilateral deafness, BAER testing was a reliable and relatively non-invasive technique to assess hearing. However, there's no way to know if a particular dog is permitted to breed until it's too late, because the hearing test is not abnormal until the dog starts to go deaf. Therefore, a genetic test will be need to diagnosis later-onset deafness of Border Collies in the near future.

Acknowledgements

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References

1. Ahlstrom L, Wilson W, Mills P. Unilateral deafness in a white Bull Terrier diagnosed by BAER assessment. Aust Vet J 2005; 83: 742-743.
2. Cox C. Investigation of hearing loss in dogs. In Pract 2002; 24: 494-501.
3. De Risio L, Lewis T, Freeman J, Stefani A, Matiasek L, Blott S. Prevalence, heritability and genetic correlations of congenital sensorineural deafness and pigmentation phenotypes in the Border Collie. Vet J 2011; 188: 286-290.
4. Igarashi M, Alford B, Cohn A, Saito R, Watanabe T. Inner ear anomalies in dogs. Ann Otol Rhinol Laryngol 1972; 81: 249-255.
5. Lynch ED, Lee MK, Morrow JE, Welcsh PL, Leon PE, King MC. Nonsyndromic deafness DFNA1 associated with mutation of a human homolog of the Drosophila gene diaphanous. Science 1997; 278: 1315-1318.
6. Platt S, Freeman J, Stefani AI, Wieczorek L, Henley W. Prevalence of unilateral and bilateral deafness in border collies and association with phenotype. J Vet Intern Med 2006; 20: 1355-1362.
7. Schraders M, Oostrik J, Huygen PLM, Strom TM, van Wijk E, Kunst HPM, Hoefsloot LH, Cremers CWRJ, Admiraal RJC, Kremer H. Mutations in PTPRQ are a cause of auto-

somal-recessive nonsyndromic hearing impairment DFNB84 and associated with vestibular dysfunction. Am J Hum Genet 2010; 86: 604-610.
8. Sims MH. Electrodiagnostic evaluation of auditory function. Vet Clin North Am Small Anim Pract 1988; 18: 913-944.
9. Strain GM. Congenital deafness and its recognition. Vet Clin North Am Small Anim Prac 1999; 29: 895-907.
10. Strain GM. Aetiology, prevalence and diagnosis of deafness in dogs and cats. Br Vet J 1996; 152: 17-36.
11. Strain GM. Congenital Deafness in Dogs and Cats. Compend Contin Educ Pract Vet 1991; 13: 245-253.
12. Strain GM, Kearney MT, Gignac IJ, Levesque DC, Nelson HJ, Tedford BL, Remsen LG. Brainstem Auditory Evoked Potential Assessment of Congenital Deafness in Dalmatians: Associations With Phenotypic Markers. J Vet Intern Med 1992; 6: 175-182.
13. Wilson WJ, Mills PC. Brainstem auditory-evoked response in dogs. Am J Vet Res 2005; 66: 2177-2187.
14. Yokoyama JS, Lam ET, Ruhe AL, Erdman CA, Robertson KR, Webb AA, Williams DC, Chang ML, Hytonen MK, Lohi H, Hamilton SP, Neff MW. Variation in genes related to cochlear biology is strongly associated with adult-onset deafness in border collies. PLos Genet 2012; 8: e1002898.

뇌간 청각유발전위 검사에 의해 진단된 보더 콜리의 양측 후발성 감각신경성난청

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요 약 : 4년령의 암컷 보더 콜리가 청각장애 평가를 위해 내원하였다. 청력 소실의 원인을 확인하기 위해 임상 검사, 신경계 검사, 검이경, 자기 공명 검사법이 실시되었지만, 특별한 이상을 확인할 수 없었다. 청력 소실을 평가하기 위해 뇌간 청각유발전위 검사가 실시되었고, 양측성 감각신경성난청을 확인하였다. 여러 검사와 병력 확인을 통해 후천적 청력 소실의 가능한 원인들을 배제하여 유전성의 양측 후발성 감각신경성난청으로 잠정 진단하였다. 이 보고는 개에서도 사람에서 보고된 유전성의 후발성 감각신경성난청이 있을 수 있다는 것을 제안한다.

주요어 : 보더 콜리, 뇌간 청각유발전위 검사, 개, 후발성 감각신경성난청