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The Genetics of Deafness in Dalmatians

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In dogs, deafness is linked to the white coat color. Approximately 85 breeds of dogs have been reported to be susceptible to congenital deafness. In Dalmatians, deafness is linked to a gene involved in their patchy baldness coloration. Dalmatians are the most prevalent with approximately half of the breed affected in the United States. A small percentage of affected dogs are deaf in both ears, the rest are deaf in just one ear. The deafness usually occurs a few weeks after the dog is born. Deafness is a lifelong condition with no treatment. Congenital deafness in dogs can also be acquired other ways, such as intrauterine infections, oxotoxic drugs, liver disorders, or other toxic exposures before birth or soon after. Inherited deafness can be caused by a multiple of possibilities.

Introduction

Congenital deafness in animals can be acquired by a number of things, such as, intrauterine infections, oxotoxic drugs, liver disorders, or other toxic exposures before or after birth, or inherited. Inherited deafness can be caused by a gene defect that is either autosomal dominant, recessive, sex-linked, mitochondrial, or may involve multiple genes. Usually the exact cause of the deafness is hard to determine unless a problem has been observed in the breed. Deafness is usually directly associated with pigmentation patterns where the presence of white patches in the hair coat increases the likelihood of deafness. The merle gene seen in breeds including Collie, Dappled Dachshund, Great Dane, and many others, and the piebald gene seen in breeds including Dalmatian, bulldog, Beagle and many others are particularly associated with deafness in dogs. Not all breeds with these genes are affected. Deafness usually develops within the first few weeks after birth while the ear canal is still closed, which usually results from the degeneration of part of the blood supply to the cochlea, which kills the nerve cells of the cochlea leaving the dog deaf.

The method of genetic transmission of deafness in dogs is not exactly known, but is associated with melanocytes. Melanocytes are cells that produce and contain pigment and can pass on pigment to other structures such as in the hairs of the skin and the iris of the eyes. Melanocytes are also vital for normal functioning of the inner ear. Blue eyes result from the absence of pigment in the iris and are common with pigment-associated deafness, but it is not always an indication of the deaf gene. In several breeds, including the Dalmatian, individuals with blue eyes are statistically more prevalent to being deaf.

In simple Mendelian genetics, each dog carries two copies of each gene, one from each parent. There are many combinations of how the deaf gene will affect generations based on whether the gene is autosomal dominant or recessive. Thus, if the deaf gene is carried as an autosomal recessive gene, the dog will need two copies of the deaf gene to be deaf. The breeding of two carriers will result an average of 25% deaf progeny, 50% hearing carriers, and 25% normal hearing progeny free of the deaf gene. The breeding of a carrier to a dog free of the gene will result in no deaf progeny, 50% carriers and 50% free of deaf gene. When breeding a deaf dog to a carrier, half of the progeny will be deaf and the other half will be carriers. Only carriers will result from breeding a recessive gene with a homozygous dominant gene.

If the deaf gene is carried as an autosomal dominant gene instead, the results will be completely different. If the deaf gene occurs at all in this situation, the dog will be deaf. The presence of two copies of the deaf gene is unlikely to occur unless two deaf dogs are bred together.

Heredity deafness can potentially result from any of several mechanisms, such as autosomal dominant, autosomal 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 the understanding of the mode of inheritance. Also, environmental factors may interact with genetic factors to trigger expression (Strain G. M., 2003).

Since piebald is a multi-gene affected gene, genetic transmission of deafness in dogs with piebald pigment genes, such as Dalmatians, is less clear. Deafness in Dalmatians does not appear to be autosomal dominant, since deaf puppies result from hearing parents. It also does not appear to be a simple recessive disorder, since unilaterally deaf and bilaterally deaf puppies have resulted from two deaf Dalmatians being bred when only deaf puppies should have been seen with this breeding. Also, there are no known reports that show deafness in dogs is X-linked to mitochondria. Thus, making it difficult to determine how deafness in Dalmatians is acquired.

Dalmatians are usually born with normal hearing, but some become deaf after a few weeks after birth. Deafness can either affect one or both ears. Dogs that are deaf in only one ear are likely to live a normal life, whereas dogs that are completely deaf are significantly handicapped. Total deafness can cause a variety of welfare problems. Dogs that are deaf in both ears can be a danger to themselves if not managed properly. Deaf dogs can be trained to follow hand signals, instead of voice commands. Deaf dogs are at higher risk of trauma, particularly from vehicles and aggressive attacks from other dogs. They may also be more difficult to train. As a result of this, many breeders have deaf puppies euthanized. Deafness is life-long. There is no cure for congenital deafness.

Recent Progress

The only way to ensure a dog has normal hearing is by testing. Mild behavioral changes may be seen in affected puppies, but some normal puppies may show poor responses to auditory stimuli and normal responses will usually be seen in dogs deaf in one ear, so casual auditory test may be of limited use when detecting affected puppies. The Brainstem Auditory Evoked Response (BAER) has been the standard test for years. Puppies are often tested around 5 weeks old, because puppies are born with a covering across the ear which prevents sound from reaching the functioning hearing organs. In normal development, this disintegrates about the same time the eyes open, so BAER can only be performed after disintegration. However, individuals can be tested at any age after this has happened. During the test, a sound is delivered to each ear and the brain response to this sound is detected using electrical sensors attached to the skin over the head. To reduce the chance of deaf puppies being born, it is recommended that only dogs that have been BAER tested and proved to be able to hear in both ears should be used for breeding. However, normal parents can produce deaf puppies, so even this does not eliminate the deaf gene completely. In return, deaf parents can produce hearing puppies. It has been noted that Dalmatians with normal hearing in both ears, tested by BAER, produce fewer affected puppies than those showing evidence of deafness (Strain, 2011).

Conclusion

Dalmatians are usually born with normal hearing, but some can become deaf a few weeks after birth. Deafness in Dalmatians can be caused by a number of things. It has been proven that there is a gene responsible for deafness in Dalmatians; however that gene is currently unknown. Dalmatians can either be unilaterally (deaf in just one ear) or bilaterally (completely) deaf or they can be completely normal (hearing). The only one to accurately test if a puppy or dog is deaf is by using the BAER test, which delivers a sound to each ear and the brain response to that sound is detected using electrical2 sensors attached to the head.

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