

## CONGENITAL DEAFNESS IN ALPACAS

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Deafness has become a well-recognized disorder in llamas and alpacas. The overall incidence of this condition in camelids is unknown, but an association between deafness and the presence of blue eyes and a white hair coat appears to exist. A relationship between deafness and hypopigmentation exists across many species (humans, dogs, and cats) and is not unique to the llama and alpaca populations. Neither the mode of inheritance nor the gene defect(s) responsible for congenital deafness are known at this time.

Hearing loss can occur secondary to nerve conductive problems in the middle ear (Otis media) or due to sensorineural injury (inflammation or degeneration of the peripheral auditory receptors or cochlear nerve). Congenital hearing loss in most species is due to premature degeneration or abnormal development of sensorineural structures. The brain stem auditory evoked response (BAER) is a sensitive method to distinguish between conductive and sensorineural hearing loss. The BAER is a rapid, non-invasive evoked potential test that does not require patient attention or any type of behavioral response to the stimulus. Ear headphones or ear inserts are used and three small needle electrodes are placed subcutaneously over different areas of the scalp. A stimulator generates clicks and the scalp electrodes record averaged electrical activity originating from ear and brain stem structures. A reproducible series of 5 waves are produced which represent sequential neural activity being relayed along the auditory pathway (Figure 1). Wave I is generated by inner ear auditory hair cells and the cochlear nerve. Relay areas in the brain stem generate waves II through V. A simple conduction problem in the middle ear will delay and attenuate waves I through V but will not usually result in their total absence. A severe sensorineural (inner ear) injury will result in absence of wave I and all subsequent waves (II through V). This is called a "flat line" response and is seen commonly in animals with congenital deafness. This "all or none response" makes the BAER a very sensitive and reliable neurodiagnostic test for congenital deafness.

Initial work on assessing normal BAERs in camelids has allowed us to fine tune the BAER protocol for the llama and alpaca species. Due to unusual external ear canal anatomy in camelids and difficulties in preventing occlusion of ear inserts by ear canal wax and debris, the headphone method of stimulation was compared to the ear insert method for BAER measurement. Results indicated that the headphone method was more reliable, yielded reproducible results, was technically easier, and did not decrease the quality of the BAER waveforms. BAER measurements were done in both sedated and non-sedated crias and adults. General anesthesia was not necessary. Young crias could be done without sedation. Older crias and adults only occasionally required sedation. Sedation was rarely required if headphones were used instead of ear inserts. The equipment was also used to assess hearing in 14 sedated and non-sedated alpacas at a private breeding facility. Good quality BAERs were obtained on all individuals in this remote location and the headphone method was very well tolerated.

Clinical studies at OSU are in progress to assess the relative incidence of deafness in alpacas and llamas with various coat and eye colors. Phenotypic characteristics that are strongly associated with and/or predictive for deafness will be statistically evaluated. Data accumulated to date indicate that deafness is strongly associated with a solid blue eye color in conjunction with a white hair coat. There are rare exceptions. With this information, breeding studies of affected and related individuals, and candidate gene analyses, the genetic basis of this trait will be better understood.

Parallel studies are focusing on characterizing early development of auditory function in crias utilizing the BAER. As congenital deafness in camelids appears to be due to premature degeneration of sensorineural structures early in life, it is important to know the normal maturation of hearing abilities in unaffected crias so that abnormal hearing during various stages of development can be recognized. To date, "normal" BAER recordings have been obtained in hearing crias as young as one day of age. Whether congenitally deaf crias are truly deaf at birth or have a delayed degeneration of auditory function after birth is not known. Further studies in this area should help define the developmental stage at which hearing abilities are lost in affected camelids. Conclusions to date based on our research:

1. A modified BAER method (using headphones instead of ear inserts) has been validated and appears superior to other methods in llamas and alpacas.
2. Hearing, as determined by the BAER, in llamas and alpacas is very well developed at birth. We are studying the BAER changes that occur during cria maturation now.
3. Congenital deafness is prevalent in camelids with a white hair coat and solid blue eye color (approximately 90 % of animals with solid white hair coats and solid sky-blue to white eyes are deaf; we have found one solid colored eyed and white coated female with deafness and one solid shy-blue eye with red coat with normal hearing). The deafness appears to be sensorineural in origin.
4. Congenital deafness was not found in llamas or alpacas with other combinations of phenotypic coat and eye color traits except for the one female with a dark eye and white coat.
5. It appears that deafness is present and can be determined immediately after birth in crias. This means that no delay is necessary to determine if a suspect cria can hear.
6. More studies are in progress to determine onset and inheritance of congenital deafness in camelids. We are trying to determine mode of inheritance and determine genetic markers for this deafness trait, which is likely to be genetic.

Research progress for congenital deafness is severely hindered by lack of funding. To date, all clinical research has been supported through the generous donations of private supporters such as: the Ohio River Valley Llama Association, A. L. Pacas Farms, the Camelid Health Foundation, and the Magical Farms Alpacas. Alpaca owners throughout the United States, including Alaska generously donated animals used in breeding research! Without further funding, this report may be as far as we are able to go to further characterize this defect on your behalf. If you wish to support this research, please contact Dr. David Anderson at 614-292-6661 or [Anderson.670@osu.edu](mailto:Anderson.670@osu.edu).

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"The Foundations, as well as several individual associations, have rejected every grant ever sent to them to fund BEW research.

Through the generosity of select breeders, such as yourself, Ray Nanko, and many others, we have managed to do some deafness research. We can not do the work we want to do with genetic markers etc because it is very expensive and the various funding agencies have not seen the research as being important enough to fund.

With what we have done, we have found that the fact that they have blue eyes actually has little to do with deafness other than the fact that it increases the likelihood that they are deaf IF they have a white hair coat. We have dispelled many myths:

1. blue eyed alpacas are always deaf - NOT true. We have tested many blue eyed but not white or only partially white animals that can hear.
2. Colored eyed alpacas can always hear. NOT true - we have tested some animals that are deaf but have colored eyes. Many have some flecks of blue or grey. Certainly most deaf animals have blue eyes, but some colored eyed animals are deaf.
3. If the skin is black they can hear no matter what the color of the eyes or hair. NOT true. Many of the deaf alpacas we have tested have black or darkly pigmented skin.

What we do know is that recessive color genes do seem linked to deafness - probably because the gene loci are close together on the DNA. Thus, if the color gene does not get "turned on", the hearing gene does not either. This is most closely tied to hair coat color, but is amplified when eye color is recessive too. Recessive colors are primarily greys with blue discoloration in eye. We have seen two grey studs where upwards of 20% of offspring have been deaf. This year we have 8 BEW females waiting to give birth. The first one born is white, has blue streaks in eye but dark pigmentation in over 50% of the eye, and is **STONE COLD DEAF!**

At some point, someone needs to decide if deafness as a birth defect is actually worth researching and fund it! I do not care who researches it, but I do think that it needs to be done."

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