

# Ask Dr. OES

A Column from the Health and Research Committee

Send your questions to oeshealth@gmail.com.

Dear Dr. OES,  
Is Cerebellar Ataxia the same as Cerebellar Degeneration? If so, why does OFA call it Cerebellar Degeneration now?

Confused in Wise, VA

Dear Confused,

Correspondence with OFA confirms that they have always called it Cerebellar Degeneration (CD).

A different genetic disease on the OFA website that may have caused the confusion is Neonatal Cerebellar Ataxia, but this only occurs in Coton de Tulear and Havanese.

Many people and labs use the terms Cerebellar Degeneration, Cerebellar Ataxia, and Cerebellar Abiotrophy interchangeably to refer to the same inherited disease. More than likely, people say Degeneration and Ataxia because it's easier to pronounce than Abiotrophy.

Cerebellar Degeneration is the deterioration of neurons in the cerebellum, the part of the brain that controls balance and coordination. CD may be inherited or result from a non-inherited condition.

Cerebellar Abiotrophy is an inherited degenerative cerebellar disease. Abiotrophy refers to spontaneous, premature neuron death, mainly occurring as a result of metabolism error.

Cerebellar Ataxia is a lack of muscle coordination caused by a dysfunction in the cerebellum.

Basically, Cerebellar Ataxia is the symptom. The specific inherited disease is Cerebellar Abiotrophy, which is a type of Cerebellar Degeneration.

Other confusing terms are Cerebral and Cerebellar. Cerebral means relating to the cerebrum, the largest part of the brain, divided into two halves. Cerebellar means pertaining to the cerebellum, the part of the brain located at the base of the skull.

*Dr. OES*

Dear Dr. OES,  
I'm confused about "cleared by parentage". What exactly does it mean? Also, I've heard recently that cleared by parentage is only good for 1 generation. Why is that?

Who's Yo Daddy in Miami, FL

Dear Daddy,

"Cleared by parentage" means that the offspring can be considered clear of a hereditary disease (without being tested) if both the sire and dam have tested "clear" for that specific disease by DNA tests. Every dog has 2 copies of each gene.



**CLEAR** means that the dog has two copies of a normal (unmutated) gene. The probability of passing down a mutation of this gene to the offspring is negligible.

**CARRIERS** have 1 normal copy and 1 mutated copy of a specific gene. The probability of passing on the mutated gene is 50%.

**AFFECTED** dogs have 2 copies of the mutated gene. They will exhibit clinical signs of the disease. The probability of passing on this particular gene to the offspring is 100%.

(Frequency of gene mutation occurring in offspring from these breeding pairs is in the chart below.)

The OFA (Orthopedic Foundation of Animals) will only certify the first generation of untested offspring from a clear sire and a clear dam if both of their test results have been registered with the OFA, and if all three dogs (sire/dam/offspring) have been DNA-identity profiled and parentage verified. The OFA certification will carry the suffix "CBP" (clear by parentage) indicating that the dog itself was not tested and that the clearance is based on the test results of the sire and dam and known inheritance patterns at the time the clearance is issued.

One reason "cleared by parentage" is only good for one generation is that mistakes are made, such as an error in the lab, but the main one is that genetic mutations appear spontaneously and unpredictably.

*Dr. OES*

|              | CLEAR SIRE  | CARRIER SIRE   | AFFECTED SIRE  |
|--------------|---|--|--|
| CLEAR DAM    | 100% of offspring will be CLEAR                                     | 50% of offspring will be CLEAR<br>50% of offspring will be CARRIERS                                      | 100% of offspring will be CARRIERS                                     |
| CARRIER DAM  | 50% of offspring will be CLEAR<br>50% of offspring will be CARRIERS | 25% of offspring will be CLEAR<br>50% of offspring will be CARRIERS<br>25% of offspring will be AFFECTED | 50% of offspring will be CARRIERS<br>50% of offspring will be AFFECTED |
| AFFECTED DAM | 100% of offspring will be CARRIERS                                  | 50% of offspring will be CARRIERS<br>50% of offspring will be AFFECTED                                   | 100% of offspring will be AFFECTED                                     |