**Breeding Strategy to Control Genetic Disorders in the Hedlund Husky Preservation Project**

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**Background:**

Prior to 2012 nearly all dogs and bitches chosen for breeding by members of the Hedlund Husky Preservation Project were selected based on a combination of athletic performance and other observable traits consistent with the line description developed by project members including size, work ethic, gait, conformation and temperament.  Very few potentially heritable diseases had been diagnosed within the line and those that did occur seemed to be easily managed. For example, in 2008 one 18 month old bitch from an outcross litter (half-Hedlund husky) was diagnosed with primary hypothyroidism and her brother had bilateral cryptorchidism (undescended testicles). They were spayed and neutered to remove them from the gene pool.

In 2009 a bitch was removed from the breeding pool due to a large number of missing teeth (10) and it was noted by the breeder/owner that dogs from some earlier related litters had some dental issues as puppies, but none so severe as to trigger alarm. However, in 2012 the presence of a single puppy in an outcross litter of Hedlund Huskies was affected with an overbite severe enough to require surgical extraction of the lower canine teeth within the first three months of life.    
  
Since then, we have had reports of Hedlund Husky puppies that that developed career-ending cases of laryngeal dysfunction similar to eosinophilic bronchopneumopathy (EBP). In 2015, a four-year-old female from a deep Hedlund Husky breeding developed zinc responsive dermatosis (ZRD), confirmed via consultation with canine dermatologists and pathodermatologists. ZRD is known to be a heritable disease in Alaskan Malamutes and Siberian Huskies, both of which have contributed to the Hedlund Husky gene pool during development of the line. In 2017 three puppies out of a litter of 4 were affected with cerebellar hypoplasia, a devastating neurological disease that affects the dog's balance and ability to walk.

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**Are the Rare Anomalies Seen in Hedlund Huskies Truly of Heritable Genetic Origin?**

When confronted with an apparently congenital defect one must determine whether that defect is caused by one or more genetic mutations. Not all abnormalities in puppies or dogs are a result of heritable genetic defects and some that are known to be genetic may also be caused by non-heritable causes. *Phenocopies* are traits known to be genetic, but that can also be produced by something in the environment. For example, cleft palate is a defect seen in nearly all breeds of dogs, but there are also 22 other causes of cleft palate including excessive vitamin A or steroidal drugs such as prednisone or prednisolone given during pregnancy

All of the following are known to cause birth defects and diseases that can mimic genetic diseases in puppies:

- Trauma (an injury caused by an outside force), including trauma while in the uterus,

injury during birth or early development.

- Bacterial or viral infections while in the uterus or acquired shortly after birth.

- Dietary insufficiencies or excesses.

- Toxins

Some genetic diseases are "multifactorial" diseases, requiring both a genetic predisposition combined with environmental components in order to express the undesirable trait. One hallmark of multifactorial diseases is the problem is identified in puppies raised in one kennel, but not seen in littermates raised in others. This may be why some puppies in the Hedlund Husky Preservation Project have had what is thought to be developmental head tremors (which they later outgrew) while some closely related puppies raised in other kennels have not. It's also possible that the laryngeal dysfunction seen in half-Hedlund Husky puppies from a single outcross litter is multifactorial.

As a general rule, if a trait is known to be inherited in sled dogs, in other breeds or in other species it should be considered inherited in our dogs unless proved otherwise. Overbite, primary hypothyroidism and bilateral cryptorchidism are known to be inherited in other lines of sled dogs as well as other breeds of dogs, so it is safest to assume that the cases we've seen within the project are heritable genetic disorders. The zinc responsive dermatosis present in one dog is definitely known to be a heritable genetic disease.

It isn’t certain that all of these disorders seen in our dogs are of genetic origin, but it also isn’t certain that they are not. Since the population of Hedlund Huskies is extremely low and consists of dogs with pedigrees that show numerous shared ancestors, members of the Hedlund Husky Preservation Project have chosen to take an aggressive approach toward ensuring minimizing the risks of genetic disorders such as those that threaten the existence of many purebred dogs to ensure that our Hedlund Huskies remain vibrant, healthy working dogs.

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**The Breeding Strategy for Control Genetic Diseases in Hedlund Huskies:**

* The Hedlund Husky is not a traditional pure breed of dog, but rather is a landrace breed. The Hedlund Husky Preservation Project does not have a "closed stud book." Our goal is not to preserve a bloodline while watching the population of dogs shrink to extinction - an inevitable result of continuous inbreeding or line breeding. Appropriate out-crossing is encouraged in order to expand the genetic diversity within our dogs. That noted, only healthy working dogs that are phenotypically and behaviorally consistent with the Hedlund Husky standard should be bred as part of the preservation program. To preserve the Hedlund Husky as a landrace, dogs selected for out-cross breedings should, if at all possible, literally be Hedlund Husky in all regards other than blood. Those out-cross candidates that do not meet the ideal standards shall be submitted to the Hedlund Husky Preservation Project Board for assessment and discussion prior to denial or approval. Once phenotypical suitability has been determined, genotype and risk of genetic disease should be assessed. Puppies from out-cross breedings should then be assessed for "type" prior to being adopted within the project.
* It is highly recommended that proposed breedings within the project be assessed for coefficient of inbreeding and kindred. Because there are gaps in the pedigrees of many Hedlund Huskies, this is most accurately done through DNA testing of both the proposed sire and dam. Both the sire and dam should also undergo genetic testing for the known genetic diseases for which testing is available.
* As has been our practice from the beginning, breeders in the Hedlund Husky Preservation Project must follow our puppies throughout their lives, seeking information from owners on at least an annual basis. Should a potentially genetic disease be diagnosed, the owners of that dog's parents and siblings must be notified and provided a copy of this breeding strategy document. In addition, project Registrar Teresa Cutler must be notified so this absolutely vital information can be included in the Project’s pedigree database.
* Any Hedlund Husky affected by a heritable genetic disease must be removed from the Project's breeding pool.  For mild disorders that can be surgically corrected or medically managed (i.e. dentition issues or hypothyroidism) this is most humanely accomplished by sterilizing the affected dog shortly after puberty or upon diagnosis of the genetic disease.  Owners of dogs affected by debilitating genetic diseases should consult with their veterinarian to determine the most humane approach to treating and/or managing the disease, taking into consideration the quality of the dog's life.

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* The presence of even a single puppy affected by an autosomal recessive or polygenic disease proves that **both** parents of that dog carry the gene or combination of genes that causes the disease.  This information should be noted in both dog’s pedigrees and provided to owners of puppies and/or owners of potential future mates. It should also be provided to the Hedlund Husky Preservation Project Registrar who maintains the database of the genetic pedigrees of all Hedlund Huskies affiliated with the project.
* Known carriers should only be bred to dogs or bitches that are known to be clear as a result of genetic testing, or who are unlikely to also be carriers based on pedigree studies.
* If a dog is affected by an autosomal recessive genetic disease, each of the affected dogs siblings have a 50% chance of being carriers of the disease. Every sibling (littermate) of an affected dog should be considered a potential carrier until proved otherwise.  Owners of those dogs should be notified, informed of the risk, and provided a copy of this breeding strategy. Potential carriers, especially littermates of an affected dog, should only be bred to dogs or bitches that unlikely to also be carriers based on pedigree studies or DNA testing.

**Rationale Behind the Strategy**

In order to control genetic diseases in sled dogs, and especially in specific lines of sled dogs, it's vital to understand that there is no such thing as a genetically perfect dog. Let's stress that statement. **There is no such thing as a genetically perfect dog.** It is estimated that every dog in the world carries at least 4 genes that, if combined with an identical gene from a mate, can cause a heritable genetic defect. While outcrossing is crucial to ensuring the genetic diversity necessary to control genetic diseases, it may also result in the introduction of other defects that haven't yet been recognized.

To understand the importance of maintaining an open stud book, please read the article "What's in the Gene Pool" at <http://www.instituteofcaninebiology.org/whats-in-the-gene-pool.html.> Historically the population of Hedlund Huskies was probably never large. Rose and Nel's Hedlund started the breed with only 3 dogs, which were then bred with other traditional village-type huskies (primarily indigenous type dogs owned by other Native Alaskan mushers) and with pure-bred Siberian huskies to establish the line. Based on the known pedigrees of today's deep-line Hedlund Huskies in the Points Unknown and Stardancer Historical Sled Dog kennels, it is estimated that the modern line of Hedlund Huskies are ancestors of only 8 'pure blood' Hedlund Huskies. Consequently, outcrossing with dogs that are phenotypically identical to Hedlund Huskies is vital to expanding the population of Hedlund's and ensuring adequate genetic diversity to prevent inbreeding depression and extinction of the Hedlund Husky.

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Fortunately, Hedlund Huskies have never been subject to the strict degree of inbreeding required of traditional dog breeds. Rose Hedlund reported to Kim Fitzgerald that she and Nels frequently refreshed the Hedlund line by out-crossing with the best Siberian Huskies they could find. Additional outcross breedings with traditional dogs from Joe Redington Sr. and from the Points Unknown Zulu line have helped maintain genetic diversity within the Hedlund Husky landrace. Recently we have even made an effort to breed dogs specifically intended to be outcrosses to the Hedlund Huskies. A breeding between Stardancer's Orion (Susan Butcher and Rusty Hagan lines) to Sirius Sled Dogs Isolde resulted in remarkably consistent puppies that are particularly similar to "pure blood" or "deep line" Hedlund Huskies.   
  
By continuing that tradition of outcrossing with suitable dogs we can help ensure the future survival of Hedlund Huskies. Nonetheless, a relatively high degree of inbreeding is necessary to preserve the traits that make Hedlund Huskies unique and special among all of the sled dogs of the world. Inbreeding results in increased uniformity of individuals within the breed or landrace and it increases their ability to pass desirable traits onto their offspring resulting in 'fixing' of desired traits and breed type.   
  
The coefficient of inbreeding and coefficient of kindred statistically measure the probability that a pair of randomly sampled genetic alleles are inherited from common ancestors. A breeding between two litter mates from parents that are completely unrelated has a coefficient of inbreeding of 25% as does a breeding between mother/son or father/daughter. From a health perspective, a lower COI is much better than one that is higher. A study of Standard Poodles discovered that dogs with a COI of less than 6.25% lived on average four years longer than those with COIs over 25%.   
  
For a more thorough understanding of coefficient of inbreeding, please read the article "A Beginner's Guide to COI" at <http://www.dogbreedhealth.com/a-beginners-guide-to-coi/>.  
  
The purpose of removing dogs affected by genetic diseases and recognizing potential carriers of such diseases requires some basic understanding of inheritance. There are four primary modes of inheritance in dogs:

Autosomal dominant

Autosomal recessive

Sex linked recessive

Polygenic and multifactorial

Each gene consists of two alleles, one inherited from the dam and the other from the sire. Autosomes are genes (chromosomes) that are not related to sex. Dogs have 38 autosomes, and 1 allosome (sex linked chromosome).

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Autosomal dominant traits are those that are expressed even though the pair of alleles are not matched. Therefore, only 1 parent need possess the dominant trait to pass it on to his or her offspring. In order for an autosomal dominant trait to be inherited 1 of the parents must also be affected. Autosomal dominant genetic diseases are easily prevented simply by removing affected dogs from the breeding pool.

Autosomal recessive traits are more complicated. Recessive traits can only be expressed when a matched pair of alleles (one of each parent) combine to form the gene responsible for the trait. Therefore *both* parents of the affected puppy must be carriers. Recessive traits tend to occur in one generation and then skip one or two generations until carrier descendants are again mated. The zinc responsive dermatosis reported in a Hedlund female is a known autosomal recessive genetic disorder that had apparently skipped several generations and thus had not been previously reported within the line.

On average, a litter from a mating between two carriers will result in about 25% of the puppies being affected, and 50% of the puppies in the litter will be carriers. If the disease is one for which no genetic test is available and there is no other way to determine which puppies may carry the recessive allele, all siblings of an affected puppy should be considered potential carriers and be bred only to dogs that are unlikely to also be carriers.

Sex-linked traits are inherited via the single allosome (sex linked chromosome) and are nearly always on the X-chromosome. On average, half of the male offspring of a carrier dam are affected with the trait. The trait may skip generations. Affected males transmit the gene to all of their daughters (rendering them carriers) and to none of the sons. If both parents are affected by the trait, all of the offspring will also be affected. For an affected female offspring to occur the dam must be at least a carrier and the sire must be affected with the trait.

It is in our favor that the heritable diseases that resulted in the overbite and laryngeal dysfunction recognized in Hedlund Huskies are probably polygenic or multifactorial traits.  Polygenic traits are controlled by two or more genes (usually by many different genes) at different loci on different chromosomes while multifactorial traits require both a genetic propensity and an environmental trigger to be expressed. Hip and elbow displasia are polygenenic traits seen in many dog breeds.

As with autosomal recessive traits, both the sire and dam must contribute the mutated genes that cause the trait to occur in the offspring.  Unlike recessive traits, however, the contribution from the sire and dam need not be equal. Because we don’t yet know the number or the specific effect of the genes involved in polygenic traits in dogs, no predictable ratios can be associated with those traits.  Polygenic traits may skip generations and may appear to be erratic in occurrence.

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By addressing polygenic diseases in the same manner as autosomal recessive traits, we believe we can eventually reduce the numbers of affected puppies produced in each generation.

**The Importance of Information Sharing Can Not Be Overstated:**

As breeders, we can not reduce the risk of perpetuating genetic disorders we don't know about. This is why it is vital that we overcome our natural hesitance to admit that genetic diseases may exist within our dogs. No one wants to admit that there might be something wrong with his or her dogs. However, we can't prevent a problem if we don't know that the problem exists.

It is vital that breeders follow up with owners of their puppies on a frequent basis to learn of potentially genetic diseased within the population, and that breeders then report that information to the Project Registrar. Whether a heritable genetic disease is recognized clinically by a veterinarian or detected through DNA testing, our Reigistrar is equipped with state-of-the art computer programs and is well educated in the field of canine genetics. These allow us to easily maintain accurate genetic pedigrees of our dogs and ensure that accurate information about them is available when contemplating potential mates within the project.

**The Role of DNA Testing within the Project.**Recent scientific discoveries in the field of canine genetics have resulted in accurate genetic tests that can determine if a dog is a carrier of a genetic mutation responsible for a many (though by no means all) heritable diseases. The list of diseases that can be prevented by using genetic tests to help make breeding decisions is rapidly growing, but is still considerably less than the 300+ genetic diseases known in dogs.   
  
While DNA testing is a very valuable tool and is becoming more so with each passing day it's important to recognize that its usefulness is nonetheless limited. For example, some mutations cause horrible genetic diseases in some breeds of dogs but are apparently harmless in others. DNA tests can also result in false negative results. For example, the test for cerebellar hypoplasia conducted by Embark Veterinary, one of the most advanced canine DNA labs in the field, is quite accurate for some breeds, but shows a "clear" result for Hedlund Huskies who are profoundly affected by the disease. By working with the geneticists at Embark we hope that an accurate test for this disease can be developed not only for our dogs, but for other types of dogs as well.

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DNA testing of both sire and dam prior to breeding allows us to reduce the risk of passing on autosomal recessive disorders for which reliable tests are available to nearly zero, but they can not prevent those for which no test is yet available those which are almost certainly present in our dogs, but have not yet been identified.

However, when we combine DNA testing while minimizing coefficients of inbreeding and kindred, we can greatly reduce the risk of producing puppies that are homozygous (affected by) any mutation. The better, albeit more expensive, DNA testing labs allow us to do this more efficiently by directly measuring the coefficient of inbreeding as opposed to making calculations based on incomplete pedigree data.

**What is the Genetic Future of the Hedlund Husky?**

The goal of the Hedlund Husky Preservation Project is to ensure that future generations of dog mushers and fanciers can enjoy the unique combination of conformation and temperament that makes the Hedlund Husky one of the most versatile and desirable sled dog and companion on the planet. To achieve that goal, we must increase the population of healthy Hedlund Huskies while minimizing the potential for genetic diseases and disorders.   
  
This can be done, and in fact we have advantages that breeders of some rare Kennel Club breeds do not enjoy. We are still able to increase genetic diversity within our dogs (no closed stud book), and the tools needed to reduce the risk of perpetuating genetic diseases.   
  
Our landrace is not the only sled dog to be in a precarious genetic situation. While we can track our line back to only 8 founders, the AKC recognized Chinook breed was founded with an effective population of only 3. While the effective population of deep-line (near pure blooded) Hedlund Huskies is only about a dozen dogs, in the early 1980s the total potentially reproductive population of Chinooks was only 11. To learn about the Chinook breed's preservation efforts, please read the article "Chinook Project" at [http://www.instituteofcaninebiology.org/chinook.html](http://www.instituteofcaninebioloty.org/chinook.html)  
  
Through careful breeding practices based on the best currently available scientific information we are confident that the line of sled dogs developed by Rose and Nels Hedlund will continue to be a presence on wilderness trails and loving homes well into the future.

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