Breed Specific Breeding Strategy

MAINE COON

Table of Contents

ATTACHMENT A – Version log

1. Introduction

The Maine Coon breed is one of the largest purebred cats, and the breed has been at the very top of the popularity list for several years in NRR (Norske Rasekattklubbers Riksforbund). Worldwide, it is also one of the most widespread breeds, based on figures from FIFe, LOOF and TICA. Originating from northeast America, the Maine Coon was initially the farm cat from the state of Maine, and known for its size and rugged demeanor, but also its friendly temperament - “the gentle giant.”

Maine Coon-ringen i Norge (The Maine Coon Breed Club of Norway), Norway's first and largest breed club for Maine Coon, decided in the fall of 2022 to create a Breed Specific Breeding Strategy for Maine Coon. The working group has consisted of Anne Marit K. Berge, Bettina Bachmann and Torhild Birkeland.

We have in this work chosen to include some in-text references, for example for source material presenting new knowledge and/or recently publications with information not widely known. Sources in general and a complete refence list can be found in chapter 7.

This first Breeding Strategy will be revised after one year, aiming to get more extensive statistics and facts for the status of the breed. Subsequently, revisions every five years will be more useful.

Any comments regarding this version can be sumbitted to ras@mainecoonringen.no.

2. General

"The Maine Coon is a natural breed of amiable character that traces its origins to the working cats found on the farms of Northeast America."
(The overall description in the breed standard of the Fédération Internationale Féline, FIFe.)

2.1 History of the breed – myths and facts

The Maine Coon is a natural breed that originated naturally from the domesticated cat population in the northeastern United States, primarily in the state of Maine. As early as the mid-1800s, it is described in the sources, and was mentioned as "the Maine Cat". The description at the time said that this cat was large, powerfully built, with semi-long fur and bushy tail.

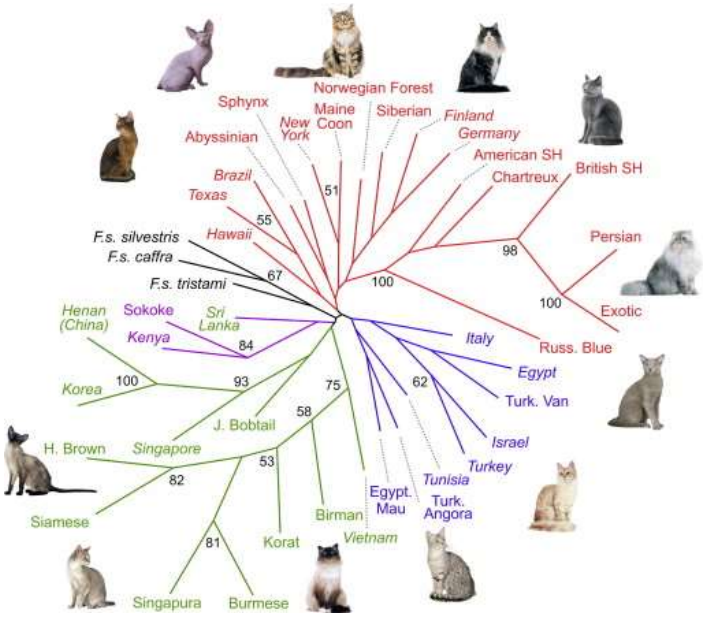


Figure 1 Lapinski (2008) used DNA to research how the different cat breeds and domestic cats are related

There are several myths and legends surrounding the breed's origins. A legend reports that the cat is a cross with the raccoon, from where it got its bushy tail, pattern, and love of water. Another story is that a ship captain named Coon was the one who brought the first cat to the United States and Maine. A third says that the breed are descendants of six longhair cats that Marie Antoinette had smuggled from France to America, in the hope that she herself would follow them.

Since there were no cats on the American continent until at some point in history Europeans brought cats across the Atlantic, we know that the Maine Coon breed originated from this first population of domestic cats that developed naturally in the northeastern United States. This population has then been mixed with cats that had been brought across the Atlantic in more recent times. Thus the origin of Maine Coon probably includes both "Angora cats" (longhair cats) and shorthair cats brought to New England by sailors, or longhaired cats brought to "Vinland" by Vikings. The cold winters of New England then made the breed, by natural selection, develop into a large cat with thick, water-repellent fur.

The Maine Coon cat's closest relatives are the domestic cats of these northeastern states, but we also know that the breed is related to the Norwegian Forest Cat and the Siberian Cat, which, like the Maine Coon, are natural breeds. By natural breeds we mean breeds that have arisen naturally from the domestic cat population in a more limited geographical area over a long period of more or less natural selection.

The first Maine cat known by name was *Captain Jenks of the Horse Marines*. The owner Mrs. E. R. Pierce wrote a book about cats and their origins, describing Captain Jenks as "One of the long-haired cats of the kind often called Maine cats... Their origins go back further than the oldest inhabitant."

When the first cat shows were held in the late 1800s and beyond, the Maine Coon was a very popular breed. Eventually, the more exotic Persian came to the United States, and interest in the domestic Maine Coon waned to the point that it almost disappeared, except at the farms of the northeastern states.

In 1953, some enthusiasts tried to revive the breed, and *The Central Maine Cat Club* was formed. The club arranged exhibitions for the Maine Coon, and the interest was renewed.

Figure 2 Cosey, Maine Coon and winner of the very first cat show in the United States May 8, 1895

Maine Coon Breeders and Fanciers Association worked in the 60s for the approval and champion status of the breed. They also prepared what is considered to be the breed's first standard. Maine Coon cats today can trace their ancestry back to the few hundred cats that were first recorded in the stud books in the 1960s and 70s, *The Foundation Cats*. These were collected throughout the United States, and some have also tracked down and received approval for more recent foundation cats from northeastern America that meet the requirements to belong to the breed.

In 1978, the Maine Coon was first shown in Europe at a cat show in the former West Germany. The first cats arrived in Sweden in 1986, and in 1994 the first litter was registered in Norway. From the almost 25 years since the breed was established in Norway until today, the breed has become by far the largest in Norway in number, with over 1,000 new kittens registered each year in the NRR.

2.1.1 Top 5 Foundation

The breed started with a few hundred foundation cats, but a few had many more descendants than others. 5 foundation cats in particular became much more significant to the breed we have today, we call these Top 5, and today's cats have about 70-75% of their ancestry from these five (percentages behind each of the cats) from the late 60s/early 70s:

- Andy Kat of Heidi Ho – about 20%
- Bridget Kat of Heidi Ho – about 20%
- Dauphin de France of Tati-Tan – about 15%
- Tatiana of Tati-Tan – about 8%
- Whittemore Smokie Joe (Smokie Joe of Whittemore) – about 7%

2.1.2 Clones

In addition to the Top 5 cats, the breed also suffered a genetic bottleneck in the 80s, which are referred to as the "clones". These clones are several offspring from the combination of Sonkey Bill and Polly Adeline that were all very similar in type, hence the term clones. The clones became very popular and line-bred over several generations, and most Maine Coons today have around 35-40% of their genetic makeup from these cats, some even more.

2.1.3 Polydactyl Maine Coon

Polydactyly was and still is a common trait in the breed. The word polydactyly comes from the Greek word polydactylos (πολύς (poly)=many; δάκτυλος (dactylos) = fingers) and describes the physical anomaly in humans and animals that have more fingers and/or toes than the *wild type* of the same species.

Polydactyl Maine Coon originally contributed to a large proportion of the gene pool in the breed (40% according to FIFe Breed Council MCO and Henning Mueller-Rech, 2011/2012). Polydactyl cats are therefore included in the pedigree of most of today's Maine Coon cats.

Polydactyly in the Maine Coon is a phenotypic variant of the breed that poses no health risk (Lange et al., 2014, Hamlin et al. 2017).

Today, one can breed and register polydactyl Maine Coon in CFA, New Zealand Cat Fancy Inc. (NZCF), The International Cat Association (TICA) and Cat Fanciers Federation (CFF). NZCF was the first feline organization to accept the polydactyl Maine Coon in the show ring on a par with other Maine Coon cats (January 2009). TICA followed this example and since 2015 has included the polydactyl Maine Coon in its shows/exhibitions. January 1, 2014 FIFe banned the breeding and registration of polydactyl Maine Coon.

2.2 Overall goal of the breed

It is an overarching goal that the Maine Coon should continue to be a breed rooted in its origins as a working cat from the farms of Maine, with its rugged character and its extremely gentle and good-natured temperament – *the gentle giant*.

3. Population of the breed

The Maine Coon breed has been Norway's most popular cat breed for several years. Worldwide, the breed is also one of the most popular, and is also by far the most numerous breed in Europe. Today, there are many more breeders in Europe than in the countries of origin USA and Canada.

For the Maine Coon, we currently have a partially open studbook, open to register novice cats from north-eastern America provided they meet the requirements of documentation and type. Crossbreeding is not allowed.

The large European population is the most dominant in the breed, with the largest number of individuals. The "pure" U.S. population based in TICA/CFA is currently less related to the European one, and is therefore valuable as outcross for the European Maine Coon. The fact that we can register and use novices means that we also have a separate "outcross population", consisting of new and unrelated foundation cats in relation to the existing population. In addition, there are also a few older lines that came to Europe before the clones, and are less related to the main European population. There are some individuals left in breeding, and often these are somewhat more moderate in type than the main population and are rarely seen in shows.

Novices, "new foundation" from northeast America provide an opportunity to make sure the gene pool doesn't get too small. There are requirements for documentation of origin; Only cats from the same geographical area of origin as the breed, in the northeastern parts of America, can be approved as foundations. The first generations, F1-F4, are experimental and are recorded in the RIEX control studbook. These cats must be monitored with care regarding what is added to the gene pool, both in terms of type, genetic disorders, general health and temperament. There are also older and newer missteps in the breed where cats from Europe and Russia have been registered as novices, which are found behind in the lines of cats today.

Since the arrival of the first Maine Coon in Norway in 1993, well over 500 Norwegian breeders have been involved in the breed. Most have bred from cats in the main European population, often called show cats, with an average total inbreeding (COI) of 15 – 20 %.

3.1 Population size in Norway

From a genetic perspective, it is not very useful to only look at the national population size , since cats in both Norway and Europe are predominantly of the same origin and thus closely related. It is nevertheless interesting to look at national population figures to be able to say something about the size of the Norwegian population.

Based on an estimate, based on the average number of new registrations in the last 5 years is 1087, we get 8697 cats registered in the last 8 years of Norway. We know little about life expectancy but if we assume that 25% of the population have died before the age of 8 we get an approximate population of **6500** NRR registered Maine Coon cats in Norway.

(Table next page)

Table 1: Number of registered cats from 2017-2021 in NRR:

Year	Number	Sire	Dam	Imports	Class 2*	Class 2%
2021	1275	675	600	49	651	51 %
2020	1362	730	632	55	741	54 %
2019	1086	587	499	80	520	48 %
2018	904	456	448	58	438	48,50 %
2017	809	388	421	59	457	56,49

**Class 2 of the NRR refers to cats that for various reasons are registered with a breeding ban by the breeder.*

There are invariably about 100 different active catteries each year registering litters in NRR (figures from MinKatt 2017-2021).

3.2 Imports

Maine Coons are imported to Norway every year. In the period of 2017-2021, 301 cats were imported by NRR members. These come mainly from Russia and countries in the European Union. A few breeders account for a large amount of these cats, while a large number of breeders account for a small number of imports. Of the total number registered in the same period, imports count for only 5.5 %. Maine Coons in Europe and Russia basically come from the same lines that we have in Norway, and thus imported cats does not necessarily mean that we get new blood or lower inbreeding in the breed here in Norway. This may explain the low percentage of imported cats. The Maine Coon population must be seen as a global population, where primarily the United States has a sub-population of the breed. There we find lines with a lower proportion of clones than in Europe, however there is also now a large number of imports even back to the US from the EU/Russia.

3.3 Average litter size

The average litter size of the Maine Coon has been about 3.9 in recent years in the NRR. Maine Coon gets litters in the size of 1-12 kittens, where more than 9 is unusual.

3.4 Degree of inbreeding

Maine Coon is a breed with very good overview of a large proportion of individuals from the origin of the breed and up to the present generations. This provides good opportunities to see which individuals recur in the pedigrees, as well as calculate inbreeding. The breed had a genetic bottleneck at the breed's founding, when the well-known "top 5" and "clones" were overused, as we can read from the history of the breed. As a result, the breed's original genetic variation was not utilized in the early years.

Breeders have access to tools such as the PawPeds database to calculate degree of inbreeding over 10 generations, as well as the complete inbreeding coefficient (COI) in the pedigree. The database is based

on voluntary submission of pedigrees from all federations and contains additional health results. There is currently a large proportion of cats from all over the world registered in the database, with 203,871 Maine Coons in the database as of 2023-02-26.

Coefficient of inbreeding COI is a measure of the proportion of a cat's locus (gene pairs) that are homozygous (identical) due to the relationship between the parents. In other words, it is a percentage value of the probability of how many loci that are homozygous, based on the parents' relationship. 100 % would indicate that the cat is homozygous for all loci.

Inbreeding should not increase by more than 0.25-0.5 percent per generation. Although the effects of inbreeding do not appear immediately, it may become apparent in later generations. A purebred cat never has 0% inbreeding, so by only looking at say 5 generations, where the inbreeding percentage can be 0%, you risk overlooking old inbreeding. New inbreeding is however more risky than old inbreeding, since inbreeding within the closest generations will accumulate homozygosity to a stronger degree. New inbreeding gives a higher risk of manifesting genetic conditions.

The inbreeding rate of most combinations made is 0% - 5% at 5-10 generations. The total inbreeding coefficient back to the foundation cats from the early 1970s is now averaging 15-20%. Due to population size, it is not difficult to avoid inbreeding in a five-generation pedigree today, but the bottlenecks from the "top 5" and "clones" should still be factors breeders today take into account in order to improve genetic diversity.

Measured heterozygosity in the breed through the WisdomPanel (Anderson 2022) screening of 7815 SNPs (base pairs) shows great variation within the breed.

This tool provides measured genetic diversity at the individual level, heterozygosity Hz. With high inbreeding, the overall heterozygosity of the entire genome (all the genes of the cat) becomes too low.

Maine Coon has a median Hz of 34.2%. Housecats has a median of 38,8 % Purebred cats overall have a median of 33.9%. Maine Coon is roughly "in the middle" and can be said to have a typical inbreeding rate for pedigree cats. The lower percentile (tenth) has a Hz of only about 30% and is too low. The upper percentile is "outcross cats" that have newer novices in the lines and thus much lower COI.

The median value is possibly "unnaturally" high due to many tested cats with newer founders within 5 generations. There is a good correlation between COI and measured heterozygosity.

S8 Table. Genetic diversity for all breeds with >15 individuals tested.

References		Median Hz	Typical Range	
Cat breeds and breed types	**>15 tested cats**	**Median**	**percentile_10**	**percentile_90**
Lykoi	104	39,1 %	31 %	41 %
Non Pedigree Cat	617	38,8 %	30 %	41 %
Cymric	16	38,3 %	35 %	39 %
Neva Masquerade	23	38,2 %	36 %	39 %
European Shorthair	91	38,1 %	35 %	40 %
Siberian	559	37,9 %	35 %	40 %
Norwegian Forest Cat	121	37,8 %	36 %	40 %
Selkirk Rex Longhair	58	37,5 %	35 %	39 %
Selkirk Rex	45	37,3 %	35 %	39 %
Selkirk Rex Shorthair	18	37,0 %	35 %	39 %
LaPerm	35	36,9 %	35 %	38 %
American Curl Shorthair	20	36,5 %	34 %	39 %
Turkish Angora	110	36,4 %	33 %	39 %
American Curl Longhair	27	35,7 %	31 %	38 %
British Longhair	43	35,5 %	30 %	38 %
Maine Coon Polydactyl	150	35,4 %	33 %	38 %
British Shorthair	347	34,9 %	31 %	38 %
American Shorthair	49	34,7 %	31 %	38 %
Sphynx	547	34,3 %	31 %	37 %
Maine Coon	**1971**	**34,3 %**	**32 %**	**37 %**
Ragdoll	1115	34,2 %	31 %	37 %
All Pedigree	10419	34,0 %	27 %	38 %
Exotic Shorthair	68	33,3 %	30 %	35 %
Bengal	1703	33,2 %	30 %	36 %
Persian	120	33,1 %	29 %	35 %
Turkish Van	40	32,6 %	29 %	36 %
Cornish Rex	106	32,4 %	29 %	34 %
Chartreux	84	31,7 %	29 %	35 %
Devon Rex	447	31,5 %	28 %	34 %
Russian Blue	64	31,1 %	25 %	35 %
Egyptian Mau	55	30,9 %	27 %	35 %
Burmilla	33	30,9 %	28 %	33 %
Somali	47	30,3 %	27 %	32 %
Ocicat	76	30,1 %	28 %	33 %
Abyssinian	167	28,5 %	27 %	31 %
Peterbald	17	26,9 %	25 %	29 %
Balinese	76	26,3 %	23 %	39 %
Oriental Longhair	51	25,8 %	24 %	28 %
Birman	174	24,9 %	23 %	27 %
Siamese	146	24,9 %	22 %	37 %
Oriental Shorthair	178	24,6 %	23 %	26 %
Burmese	113	21,5 %	19 %	27 %
Singapura	39	20,3 %	16 %	24 %
Korat	51	20,0 %	18 %	22 %

COI is a statistical measure of homozygosity, while genetic diversity shown in the table here is a measured level of heterozygosity. The lower the COI (homozygosity), the lower the inbreeding - while for measured genetic diversity (heterozygosity) it is the opposite, the higher the number, the lower the inbreeding.

3.5 Popular Sires

For dogs, NKK (Norsk kennel klubb) recommends that a single sire accounts for no more than 2% of the total number of registered puppies of the breed in the last 5 years, for numerically large breeds. In comparison, 2017-2021 is considered our last 5-year period. A total of 5436 MCO were registered during the period. Consequentially the maximum recommended number of offspring per sire is 109.

Some studs in recent times have over 150 offspring. This is well above the recommended limit but is not common practice in the breed. Most studs' sire 1-5 litters. Recommendations are to use several individuals in breeding and avoid overusing popular sires, "matador breeding". Popular sires will have far too much influence on future generations. It is therefore especially important that male cats with many offspring meet health requirements and have good type.

There is a tradition in cat breeding to use the same males with all the females in the cattery, before replacing him with a new one. Typically, you then keep daughters from your stud and buy a new male for them. In Norway the typical cattery is small in the breeder's own home, and there are challenges associated with keeping studs like that, that have probably led to this way of breeding. It is often not possible to have more than one male cat per household, and often they can start spraying indoors. Male cats rarely get together if they don't live alone with each other. It is also often difficult to borrow studs from others, which has caused this way of breeding.

3.6 Males and females used in breeding

No data are available on the distribution of male and female cats used in breeding. What we can see is that about half of registered cats annually are registered in class 1 (breeding class). A relatively even number of males and females are born each year. It is reasonable to believe that far more female cats than male cats are used in breeding. Most breeders have more queens than studs in their breeding program.

3.7 Objectives and strategy

Short-term goals:

- Limit the use of individuals (popular sires).
    - Rather use siblings of popular individuals
    - Using individuals a few times each
- Avoid incest and close matings. There are no rules at present, but incest mating should possibly be banned, and other close matings such as uncle/niece etc. should be avoided.
    - 0-5 minimize inbreeding in the closest generations
- Increase the use of genetic tests where genetic diversity is measured, e.g. MyCatDNA.
- Encourage the use of PawPeds, which is a good tool for showing the total inbreeding COI.
- Training and education about Breed Specific Breeding Stategy
- Cooperate with other breeding organizations. The principles of genetic diversity are independent of species/breeds.

Long-term goals:

- Increase the genetic diversity of the breed so that it continues to remain a healthy breed prepared for the future.
    - Using cats with lower COI in breeding

4. Health

Maine Coons are generally no more prone to health problems than most other cat breeds. However, as with most breeds bred in a closed population, the Maine Coon also has some more breed-specific health challenges.

Breeding problems that are directly related to health can be classified mainly as two types:

1. Anatomy defects due to extreme type breeding, i.e. breeding for extreme exterior traits. Such errors can be solved by selecting healthier and more natural anatomy if there is a wide enough gene pool from which to select. (See also section 5.3 Exaggerated exterior features)
2. High frequency of specific disease. This can be solved through screening programs/health programs that provide a basis for selection on healthy individuals if there is a broad enough gene pool from which to select.

If the gene pool is too small or disease/anatomy defects are too widespread, then there is no room to select too hard as this will further narrow the gene pool, with consequences such as even higher disease problems and inbreeding depression. If, on the other hand, you have a broad enough gene pool, and the opportunity to add new blood to some extent, you have the opportunity to breed away from health problems through selection.

The general health requirements in Fife's BRR (Breeding- and registration rules) apply to all breeds, and under the Norwegian Animal Welfare Act we are not allowed to breed animals that are not healthy. The selection of breeding animals must thus focus on health, and most importantly, see the cat as a whole.

The independent foundation PawPeds in Sweden is the administrator of two important health programs, HCM and HD. It links veterinary-verified screenings to the studbook, which includes most Maine Coon lines from all possible federations, and is worldwide. Results there are thus very important since continuity through the generations far beyond FIFe and NRR is detectable.

Prevalence of diseases/health problems in MCO - overview

- HCM: 5-15 %
- HD: 25-37 %
- Rarer and less serious genetic diseases: 0-5 %
- Entropion: no data exists
- Juvenile Gingivitis: no data exists
- Caesarean section: unclear

4.1 Inbreeding and hereditary disease

Most disease mutations are recessive. This means that as long as you do not mate two carriers of the same mutation, there is no problem. It is estimated that an individual at any given time will have several thousand possible recessive harmful mutations in his or her genome, and new ones may arise with each generation. This is the main reason why we do not want to engage in inbreeding or close matings, since it will cause the unknown harmful mutations to double. High diversity in a breed means less risk of such doublings, if one does not mate too closely.

4.2 HCM – hypertrophic cardiomyopathy

HCM is present in all cats, including domestic cats, with a prevalence of approximately 10% (5-15%). It is the most common heart condition in cats.

The Maine Coon breed has been featured in many more research studies on HCM than any other breed. In 2005, a colony of strongly related Maine Coons with HCM was studied to understand more about the disease, and the one harmful mutation we know of, A31P in the MyBPC3 gene, was discovered there. This

does not mean that HCM only exists in MCO, but we know much more about prevalence and cause here than for other breeds that are also at risk.

Diagnosis and symptoms:
The Maine Coon has a health program against HCM through the independent foundation PawPeds since 2004. Hundreds of veterinarians with degrees in cardiology worldwide participate in the health program and submit the results of echocardiography of the cats, and all results are published in the Maine Coon pedigree database. As of 1 June 2023, there are nine PawPeds-affiliated cardiologists in Norway.

Hypertrophic cardiomyopathy means that the heart walls thicken, making the heart inefficient. The heart may then stop, or the cat may develop blood clots. Cats with HCM can have varying degrees of symptoms, and many have no visible symptoms easy to detect. For cats that get a severe form of the disease, this leads to premature death. In other cases, the disease can develop gradually and have a relatively mild course that can be treated, and some cats may have HCM without noticeable symptoms until very old age. Diagnosis must be made through ultrasound examination, echocardiogram of the heart performed by a cardiologist, or post-mortem as an autopsy with histopathology – tissue sample examined under a microscope. A symptom often thought to be a sign of HCM is a heart murmur. Studies show that this applies to only half of HCM affected cats, and half of cats with murmurs have other, often harmless causes.

Prevalence of disease thus increases with age. For the cats examined with ultrasound, the changes in the heart will appear by the time the cats are 5 years old (Follby 2022). Cats with a normal heart at 5 years' age will only very rarely develop HCM, and then often as secondary HCM due to other diseases.

4.2.1 Health surveys

The advice in FiFe and in the health program PawPeds is to heart examine the breeding cats:

Before breeding:
Cardiac examination with ultrasound before breeding will detect early and severe heart disease, as well as other heart conditions, and should be mandatory for all purebred cats before breeding. The early examination can capture the few who have a very early onset of the disease. These have often been cats that are homozygous for HCM1-A31P, which we avoid altogether in NRR after the mandatory test requirement.

After the age of 5:
Continued cardiac examination especially of cats over 5 years old is even more important, for two reasons:

1. A good number of cats will have HCM as a *subclinical* disease, i.e. have no noticeable symptoms themselves. An ultrasound examination will show thickening of the heart, and the cat will be diagnosed even if it appears to be healthy. These cats can live about as long as cats without HCM (Trehiou-Sechi 2012). Such a cat can pass the disease on to offspring, which can acquire the more severe form of HCM, apparently from healthy parents.

2. The usual age of onset for diagnosis in cats that develop HCM is 4.5 years, shown in studies. This means that the early study will only capture the few who have a very early onset. It is essential that a new heart examination of breeding cats is performed after they have reached the age of 5, in order to include those who have the onset of the disease at a typical age. The most common is first diagnose when the cat is 4.5 to 5 years old (Follby 2022).

A cardiac ultrasound shows the status of the heart as of the date and age the cat has on the day of the examination. It is therefore important to continue HCM examinations as your cat gets older, and it shows the importance of re-examining at advanced age. Some critics claim that a cardiac ultrasound is just "a

snapshot", but it shows that they have not understood how HCM works. It's more accurate to say "still healthy heart at this age" – and the older your cat, the more important the outcome.

4.2.2 Statistics on HCM

PawPed's HCM health database contains from 2004 and as of Dec. 31. Dec. 2022 a total of 20,682 HCM examined Maine Coon cats. Of these, for the 8 Norwegian cardiologists, there are only 663 HCM examinations of MCO in total 2004-2022. 173 of the examinations were done before 31 January. 2015, and 272 are from before 2018, i.e. there are "only" 391 examinations in the last 5 years.

We see from the research (Follby 2022) that the most common age of onset for an HCM diagnosis is 4-5 years. Many cats also have subclinical HCM, which shows no symptoms (Trehiou-Sechi 2012) and can only be seen via ultrasound, but still has hereditary risk that can be transmitted to offspring. This means that repeated ultrasound examinations, even after the age of 5 years, are important to detect HCM cases and risk lines.

Of the 663 HCM tests performed by Norwegian cardiologists, 401 cats have only been scanned once. 78 cats are checked 2 times, 18 are checked 3 times, and 12 are checked 4 times. Only about 40 cats have been heart examined over 5 years old. Some Norwegian cats may also be examined by cardiologists who are not yet affiliated with the health programme, by cardiologists abroad, or by a regular veterinarian with ultrasound equipment.

4.2.3 DNA testing of HCM

There is one known genetic factor that may cause HCM in Maine Coon, namely the A31P mutation in the MyBPC3 gene, often called HCM1 DNA, discovered in 2005. Unlike most disease mutations, it is considered dominant, with variable expression. For MCO breeders in NRR, it is mandatory for breeding cats to have known status of this mutation and never mate two heterozygous cats (N/A31P HCM1). The status of cats for A31P is recorded in the studbook, shown in the pedigree, and is publically available in MinKatt. Cats that have a doubling of this mutation are at very high risk of becoming ill, estimated penetrance is about 80% or more. A slightly elevated risk is also seen in those who have one copy of the mutation (heterozygous), but these cats are normally healthy until 4-5 years of age, and the few that become ill may have a milder variant of the disease.

When A31P was discovered in 2005, it was found that 20-40% of all MCOs had this mutation. The prevalence of HCM1-A31P DNA is below 1% as of 2022 (Anderson 2022). We see today that very few cats have this mutation, and that HCM cases for MCO registered in the NRR must have a different genetic cause.

Research is ongoing in research institutions around the world to trace more genetic causes, including in other breeds. In humans, hundreds of different mutations are known in several genes for heart function that can cause HCM. Research into parallel genes in cats is ongoing to look for the causes. It is important that newly discovered mutations are validated against actual disease occurrence before it is relevant to recommend DNA testing and any mandatory tests.

Research as of 1 January 2023: Shortly after Meurs discovered A31P, Danish cardiologist Koch saw another gene variant in the same gene MyBPRC3 could also possibly be the cause: HCM2-A74T. This has not been validated since then, and this variant is now considered harmless. In 2020, McNamara published the discovery of TNNT2, a mutation in the troponin gene that can cause HCM in humans. The study there concerned a single HCM-affected MCO. Schipper 2022 examined many cats with and without an HCM diagnosis. The gene frequency for TNNT2 was determined to be 32% based on the 160 cats that have been genetically tested, it is thus very common. The 99Lives project (Leslie Lyons) showed that the mutation also exists in other breeds and domestic cats. Then 31 Maine Coons with an HCM diagnosis, and 58 healthy as control, were checked to see if there was a difference in TNNT2 status between them. The results showed

far too weak a correlation to say anything about efficacy, and the conclusion was that TNNT2 has "unknown significance" for the development of HCM in cats. This means that there are no indications to test or take cats with this variant out of breeding.

For cats with several offspring in breeding, a later examination should also be carried out, at about 7-9 years of age. This also applies to neutered, rehomed cats, which the breeder should take responsibility for collecting and examining.

All Norwegian veterinarians specializing in cardiology (ECVIM, ESAVS) should be encouraged to join the health program.

4.3 HD – Hip dysplasia

Statistics from both OFA and PawPeds show that Maine Coon has increased prevalence of hip dysplasia – HD. That is why a health program has been devised to reduce HD in the breed. X-raying of the hips before using a cat for breeding is recommended by both FIFe and the health program PawPeds.

What is HD?

The defect consists in the fact that the hip socket and femoral head do not fit each other. In cats, it is especially the hip socket that is too shallow (Keller 1999), so that the femoral head, does not fit as it should. This leads to wear of the joint. Parts of the cartilage can degenerate and get replaced by bone tissue in the body's attempt to repair the damage that has occurred. Symptoms and ailments often do not appear until the cat is older and calcifications develop in the joints. This means that problems first arise with time, and will have varying severity depending on how much calcification occurs. Cats are effective at hiding pain, so a cat with HD may not have visible symptoms. X-ray is needed to make a diagnosis.

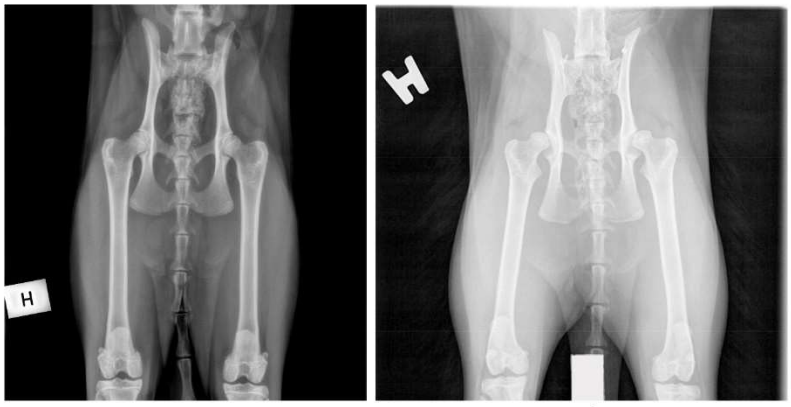


Figure 3 The images show a normal hip and a hip with HD grade 3 on both sides

HD is congenital and hereditary, polygenic, and recessive. This means that there may be a large number of genes and possible harmful mutations that are the cause, and it will not be possible to DNA test for single mutations to avoid risk. With many recessive genes, the risk of doubling increases with higher inbreeding. HD is also found in other breeds, and some point out that size may be one reason. Figures from OFA and PawPeds show that one of the breeds with a possibly high prevalence of HD is DRX, indicating that a small gene pool is a greater risk factor than physical size. HD is affected by several interacting genes, and inheritance can be masked, the defect does not have to be visible in the individual, it is enough that the cat has inherited a predisposition from both parents. So, two cats with normal hips can have offspring with hip dysplasia, and cats with hip defects can also have offspring with normal hips.

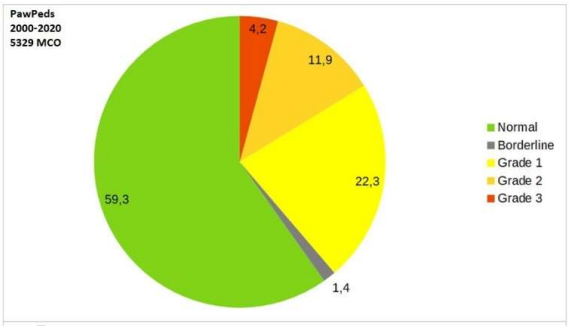
It is estimated that about 8-10% of all Maine Coons have HD of such a high degree that breeding is not recommended.

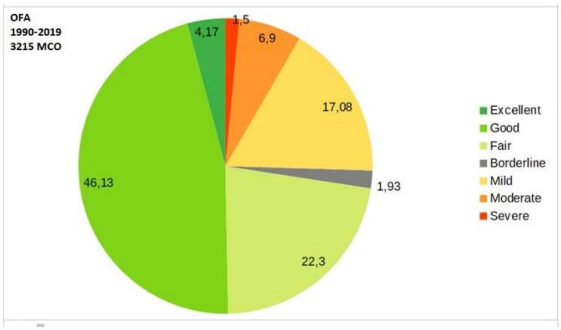
Radiographs of the cat can be performed by a veterinarian with appropriate X-ray equipment. When X-rayed according to the program's procedure, the examination form is completed and signed by owner and veterinarian. The images are sent digitally via the website from the veterinarian to the health program's reader, Elisabeth Ball at SLU in Uppsala, Sweden. The hips are ranked on a 4-part scale. The answer is normally sent to the breeder within 2-5 weeks. The results are entered in PawPed's health database, and

after 2-3 months they are also entered in the public pedigree database where everyone can look up the cat and the results themselves. It is recommended that cats are X-rayed when they are over 10 months old and within about 2 years, to get the correct result.

An alternative to PawPed's HD health program is to send the X-rays to OFA (The Orthopedic Foundation for Animals) in the United States. They use a 7-part scale and provide a preliminary assessment of cats between 12 and 24 months, read by orthopedic surgeon G.G. Keller. For cats that receive Excellent, Good (OK for breeding) or Moderate and Severe (not for breeding) in a preliminary assessment, the evaluation can be considered accurate and final (Loder 2017). For Preliminary Fair, Borderline and Mild, OFA recommends to wait with breeding and take new X-rays after the age of 24 months. Then the images are judged by a panel of 3 affiliated orthopedists.

The procedure for X-rays is the same for PawPeds and OFA, you fill in the form and take it to the vet for the radiography. The veterinarian signs that the ID has been checked and submits the form and images to the reader. OFA also provides the opportunity for veterinarians to register for direct submission of the images digitally, making everything go faster. If you get the X-rays evaluated at OFA, confirm that you accept the results to be published on their website, which does not happen automatically for cats under 24 months or for results that are poor.





Recommendation is to choose a veterinarian who has newer digital X-ray equipment, who has experience with HD X-rays and preferably has attended courses under the auspices of NKK and the veterinary association. In these courses, correct positioning (the same for dogs and cats) is taught, and ideal device tuning for the best possible images. In addition, procedures for secure identification and submission of the images are reviewed.

Evaluating HD X-rays should definitely be done by an orthopedic surgeon who specializes in HD in cats, and we recommend PawPeds, with OFA as a good alternative.

Large studies of HD in over 5000 Maine Coons show that prevalence is 37.6% (Low 2019). OFA's statistics of over 3,000 show prevalence of 25% from OFA, but there may be bias away from the most serious cases as submission is voluntary and can be stopped after X-rays have been taken. The most serious cases will be visible on the X-rays there and then.

4.3.1 Challenges

Since two breeding cats that have normal hips can have offspring with HD, many people think that there are no main genetic causes of HD. Low's study shows that heritability is 0.36, meaning that 36% of the variation for HD within the breed can be attributed to heredity. Figures from PawPeds (Low 2019) where cats have been X-rayed since 2000 and have many lines X-rayed for several generations, show that if you continue to select on normal hips over several generations, exclude serious cases in breeding, and mate cats with mild/grade 1 only with normal, then you reduce the risk for HD in offspring (Sprenger 2020).

Radiological assessment of X-rays is always a subjective assessment, so the results will never be exact and absolute. A study with parallel assessment of 196 HD images by 3 readers (Ball 2022) shows that there is generally enough consensus for the results to be used as guidelines in breeding. In the study, the X-rays were assessed twice by each reader, several weeks apart. Equal assessment the first and second time was

made in 65-71% (same reading) of cases. The differences in reading mostly go up or down one degree. The highest consensus is hips rated to grade 3, followed by normal hips. The most common change (30%) between readings is from grade 2 to grade 1. The rarest are changes of two degrees (only a few), and none were changed three degrees. It also turned out that the hips readers spent a long time assessing, were also those that had the least correspondence in the readings. This may indicate that it applied to hips in the borderland between degrees and which were more difficult to judge.

The conclusion is that the assessments have good consensus, but that some of the readings may vary by one degree up or down, and this mainly applies to hips in the middle layer between normal and severe degrees. This is in the nature of radiographic assessments and should not be interpreted as if HD reading produces completely random results (Ball 2022).

No breeding restrictions should be introduced at present, other than those that are already implicit in the animal welfare requirement that breeding animals be healthy. There are several causes for this: The evaluations may have a subjective factor, and the uncertainty is greatest for the cats that have minor abnormalities in the hip joint (Ball 2022). The combinations made have a lot to say for risk in offspring (Low 2019, Sprenger/PawPeds 2022). Narrowing the gene pool may have a greater negative effect on health in the long term than taking individual cats out of breeding, and HD prevalence is too high to breed only cats with normal hips.

Many cats with HD grade 2 and 3 have no symptoms and have few or no problems until osteoarthritis develops as they age. HD X-rays with registration of all results will raise awareness among breeders, and assessment of combinations will to a greater extent be done according to the recommendations to reduce incidence. However, a female cat with HD grade 2 and 3, or Moderate, Severe (OFA) should not be mated, as the strain on the hips during pregnancy will be great.

4.4 Ocular disorders

The Maine Coon is no more prone to eye disease than other breeds. However the condition *primary entropion* can be linked to extreme type breeding: Entropion is a disorder in which the cat's eyelid rolls in towards the cornea of the eye. Eye hair scratches the eye, causing irritation and pain. The disorder must be treated surgically.

Entropion needs surgery. If the degree is moderate, then it will not be noticeable after surgery that the cat has been treated. A more severe degree will be fully noticeable afterwards. Entropion can be primary (hereditary) or secondary (acquired). Secondary entropion can occur after injury to the eye, or inflammation and infection. Secondary entropion may also require surgical treatment.

The cause of primary entropion is about the anatomy of the cat head. Deviations from the Maine Coon standard with e.g. too pronounced muzzle, extreme cheekbones, narrow head, and deep set eyes can increase the risk for entropion. It is a matter of inheritance, and no single mutation to test for. The breeder must have knowledge of the anatomy of the head, and not breed for too extreme type. The risk is higher for male cats. Anatomy that increases risk is also not correct by the Maine Coon standard.

Primary entropion is a hereditary disease, but based on quantitative genetics, where a trait is inherited based on many different genes that work together (Kjetså 2017). Extreme breeding with focus on more extreme type can result in cats with entropion. A cat at risk should be mated to a cat without the same risks.

FIFe's standard for Maine Coon says this about eyes:

Eyes: Large and widely set.
Slightly oval, but not almond shaped, appear round when open.
Set slightly slanted towards the outer base of the ear.

Any colour is permitted. There is no relationship between eye- and coat colour. Clear eye colour is desirable.
Faults: Slanted, almond shaped

According to FIFe's rules, it is not allowed to breed cats that have or have had primary entropion.

4.5 Teeth and gums

Juvenile gingivitis is considered to have breed-specific prevalence and is more common in MCO than other breeds (Ruhnau 2017). Typically, this is a gum disease where symptoms fade as the cat matures, and most cats have few or no symptoms by the time they are 1.5-2 years old. However, some cats develop chronic ailments and severe symptoms, and lines with frequent or severe occurrence should be stopped from breeding.

Problems with teeth and dental disease are generally common for all cats regardless of breed. Cats with serious chronic problems should not be bred, and lines where the same problems recur should be considered stopped. For a cat with a known problem, only a mating against a partner who comes from lines without the same problem should be considered.

4.6 Rarer genetic conditions

There are a large number of genetic conditions that can be DNA tested, and the development of such tests is progressing rapidly, so new variants are constantly emerging that in theory can cause disease or problems. We would generally caution against over-interpreting such DNA results.

Essentially, all of these are recessive (the HCM mutations are a rare exception) and cannot cause disease unless doubled. It is a much greater danger for a breed that cats are removed unnecessarily from breeding and thereby reduce the gene pool, than for some offspring having doubled a rare mutation that, for example, gives a somewhat slower tendency to clotting, or perhaps may increase the risk of urinary stones. DNA tests should be a tool to optimize combinations in breeding, but one must not overreact and take carriers out of breeding.

4.7 A brief overview of testable recessive mutations:

Today, we have the opportunity to relatively easily and inexpensively test for a variety of mutations. DNA tests often come in packages, where you get results for a number of mutations from a single DNA sample. Not all such tests are necessarily equally relevant, but here are some individual mutations that may occur in the Maine Coon breed while not considered a major health problem.

<u>SMA</u>
Breed specific SMA. Spinal muscular atrophy is a very serious disease, but the deletion (gene mutation) that causes it is very rare and occurs in principle only in known lines. Cats in these lines where parents are untested should be checked before breeding. The risk of the mutation doubling and causing disease is otherwise minimal, and there is no need for mandatory testing or a separate health program for this. For the few lines we have, we would encourage breeders to register carrier status in MinKatt and PawPeds, and to inform others who have offspring in the lines.

<u>PK-deficiency</u>
Deficiency of the enzyme pyruvate kinase will cause anemia, to a mild to severe degree. The mutation was first detected in Abyssinians, where it has long been known to cause disease. It turns out that it has varying and fuzzy efficacy for Maine Coon. "PK-def shows significant clinical variation, including when the disease onsets and how severe it is." WisdomPanel had veterinarians examine homozygous positive MCO and has spoken to owners: "Our veterinarians have interviewed owners of ten Maine Coons [...] In three

cases, the owners reported events with at least one mild potential episode with clinical signs such as lethargy, anorexia, weight loss and/or jaundice. [...] and a female cat that could potentially have shown mild symptoms soon after birth" (Anderson 2022). There is great uncertainty about symptom expression, so one must regard the effect of PK-def as somewhat unclear for Maine Coon. As long as we do not know for sure whether PK-def causes disease and problems, we should not mate two carriers.

Cystinuria

A mutation has been detected that causes an increased tendency to crystals in the urine, which is painful for cats. Type B cystinuria is present in less than 1% of MCO and must be considered very rare.

Factor XII deficiency

Factor XII deficiency is a fairly recent disorder that is prevalent in almost all cat breeds, including about 10% of MCO tested. There are two different mutations, both very common, but they are not considered to cause severe disease. Cats that are homozygous positive lack an enzyme and will have a prolonged clotting time of the blood, but not so that transfusions are needed. There is no reason to take carriers out of breeding, but if we have a heterozygous carrier, then the best thing to do is to mate with a negative one.

MDR1 drug sensitivity

The newly discovered mutation MDR1 shows vulnerability with overreaction to some drugs, including parasiticides. Approximately 5-6% of MCO are carriers of this mutation. For a healthy cat, substances in the blood are protected from entering the brain, but cats with MDR1 defect may experience increased neurological symptoms from, among other things, parasiticides containing ivermectin or eprinomectin. As MDR1 is newly discovered and few cats have been tested, it is recommended that information about carriers be communicated to others who have cats in the same line, so they can get tested.

4.8 Blood type B

Blood type B is not a disease, but a queen's blood type B can cause kittens to die. There are 3 known b-alleles that can give B blood, and a variant c that can give AB blood. B1 is relatively common in MCO, but cats must inherit two copies, i.e. from both parents, to become blood group B. Type b2 (Siberian, Turkish breeds), and b3 (found only in Ragdoll) are rarer and not known in MCO. However, the previous study by the Laboklin team (Kehl 2019) has found two MCO carriers of c – but we can establish that it is extremely rare in MCO. It is recommended that all breeders of Maine Coon know their cat's blood group status and that the DNA results are uploaded to MinKatt.

4.9 White cats and deafness

Dominant white cats have an increased risk of deafness, and blue-eyed white cats are at greatest risk. A small study (David 2014) shows that dominant white cats that also carry the dominant white spotting allele (genotype W/Ws) have a higher risk of deafness than white cats that do not (genotype W/w). Cats with a lot of white and small amount of color can also become deaf. In FIFe, it is not allowed to mate two white cats with each other.

White cats are prone to deafness due to a mutation in a gene called KIT. There are two known mutations in the KIT gene (alleles) that each give their own phenotype. One mutation (allele) gives dominant solid white (W) while the other mutation gives the phenotype that is called white spotting and produces varying amounts of white (Ws).

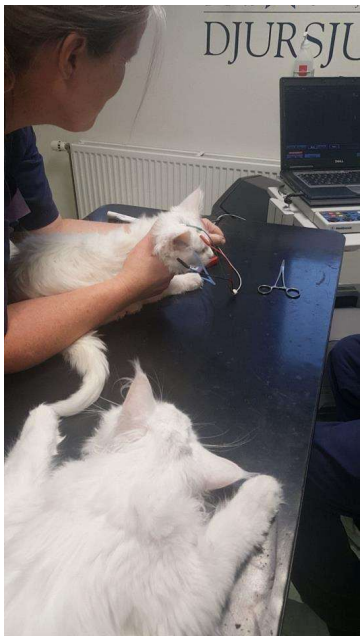
The white spotting phenotype is often referred to as bicolor, van or harlequin by cat breeders, based on the amount of white on the cat. Dominant white spotting should not be confused with recessive and spontaneous small white spots, such as lockets, which do not have the same mutations in KIT.

KIT and the KITGN genes have an influence on melanocytes. Melanocytes are cells that develop from melanoblasts, migrate in the body during early fetal development (prenatal stages) and then develop into

what we call pigment/melanin (Grichnik, 2006). These cells contribute to normal development of the inner ear in cats (Reissmann & Ludwig, 2013). KIT and the KITG gene affect melanocytes in several ways.

It can affect:
- Migration
- Production (synthesis)
- Development and survival.

There are also other genes that affect melanocytes. If the melanocytes do not reach the inner ear (cochlea) during fetal development, it may result in an overgrowth or collapse of a membrane, thus blocking nerve signals. There may also be other disturbances in the nerve signals, resulting in neural impulses not reaching the brain. This leads to deafness.



BAER stands for Brainstem auditory evoked response, and is a method designed to measure whether neural impulses travel normally through the ears, reach to, and can be interpreted as sound, in the brain. This is the only reliable method at present to determine whether a cat is deaf or hearing. It is recommended to BAER test all white breeding cats so that all white cats used in breeding are normally hearing. This will reduce the risk of having deaf kittens. In Norway, BAER testing has not been available until very recently, and it is only available in Eastern Norway. A gradual introduction of mandatory BAER testing, over some time, would therefore be recommended.

The eye color in white cats is a result of the amount of pigmentation in the eye. Low amount of pigmentation causes blue eyes, and more pigmentation gives yellow eyes. Therefore, blue eye color is also associated with a higher risk of deafness than yellow eye color. There are several other blue-eyed cats that have different genetics, e.g. Birman and Siamese. This is not relevant to the breed, so we will not go further into this here.

The study that exists shows that dominant white W- and cats with white spotting Ws should not be mated since cats with genotype W/Ws have a higher risk of deafness than Ww. Despite little research, this is something that has been known in many aquaculture environments "forever", but it appears that the KIT gene and heredity/expression have some irregular expressions.

We see that W-Ws combinations can have variable expression, as can the "gloves" mutation in Birman, which is also found in other breeds and can cause small white spots, such as a locket, "bikini" spot, or a single white toe.

W-locus and EMS: Today, EMS code 09 is used for two completely different phenomena: heterozygous dominant white spotting where less than 25% of the cat is white, but which in expression always gives white on all 4 paws, white bib, and belly button. This variant has less white than 03 (25-50%) and could benefit from using EMS code 04 "mitted", which is currently only used on RAG. EMS code 09 is also used for small white spots that are inherited recessively or arise spontaneously (Lyons 2013), and where the cat has at most a little white on a paw, a locket on the chest and/or a spot on the belly (so-called "bikini"). A current EMS code for small spots could be 08, requiring a maximum of 3 separate white spots, max one paw. Both genetic and phenotypic, these are two completely different phenomena, and it would be a great advantage if they had different EMS codes. It would then also be easier to write a breeding recommendation that all-white EMS w should not be mated to dominant white spotting 01, 02, 03 and "04" - while mating with "08" does not increase the risk of deafness.

4.10 Reproductive health

At present, we have no data on the reproductive health of the breed, but there is no reason to assume that the Maine Coon is particularly vulnerable when it comes to, for example, pyometra or caesarean sections.

4.11 Life expectancy and causes of death

Few data are known on normal life expectancy for Maine Coon, or common causes of death. Two studies based on self-report forms include a few hundred MCOs.

In Follby's study (2022), 1113 cats from several breeds are included, including 223 MCO. The most common causes of death in the study:

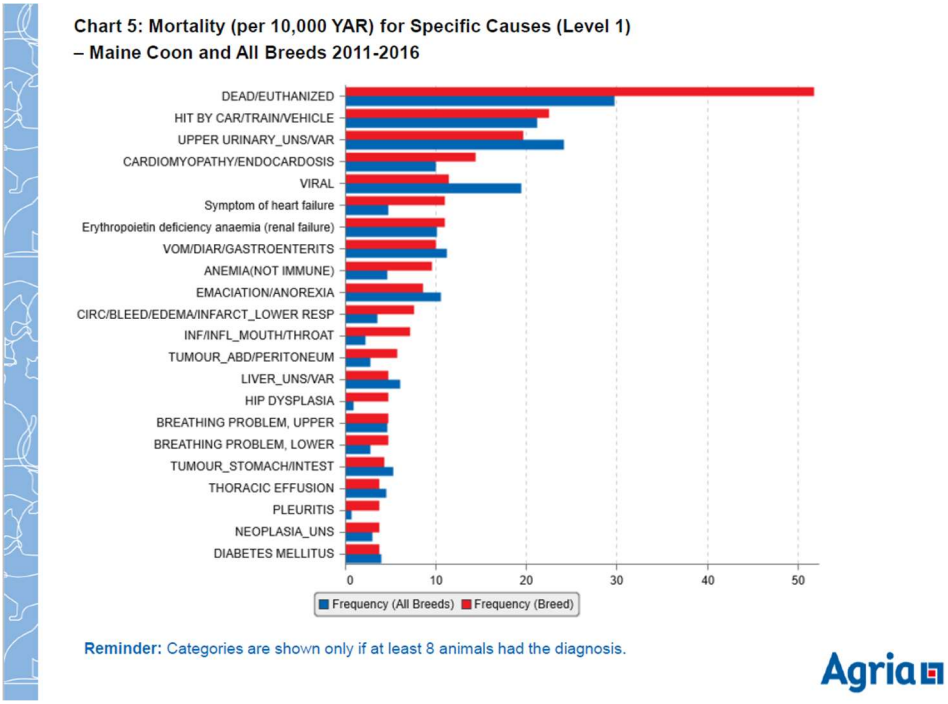
- Urine/kidney 17.6%
- Cardiac 16.3%
- Tumours 15.1 %
- Infectious disease 11.2 %

Follby shows that 1076 of the 1113 cats had information related to life expectancy that showed:

- Cats with heart disease FCM (HCM (hypertrophic) but also DCM (dilated) and RCM (restrictive) median life expectancy 9.2 years
- Cats screening result 'normal' median 13.4 years
- Cats with screening result 'equivocal' (31 cats) life expectancy median 12.3 years

Follby also points out the distinction between the results they arrived at and disease statistics from insurance companies: Teeth and joint diseases are underreported in insurance companies overviews, presumably due to terms and coverage.

Agria's statistics on causes of death in Maine Coon, compared to other breeds, for the period 2011-2016 (sample is from Sweden, Norway, Denmark, Finland and UK):



In Chapter 3.1 Population Size we have already mentioned that causes of death are not well known, and in our estimates we assumed 25% had died before 8 years of age. Life expectancy is often estimated at around 12 years looking at various internet publications, but we have not been able to find the sources for this claim. Therefore it would be desirable if cause of death and age at death could be registered in the pedigree book, to get a better basis for describing average life expectancy and causes of death.

4.12 Objectives and strategy

Short-term goals:

• Get more people to record all health results publicly, as well as date of death/cause of death. Encourage autopsy at unknown cause of death.

• Facilitate more detailed registration of health data in NRR pedigree register.

- Encourage HCM scans both before breeding and after 5 years
- Data for caesarean sections/stillbirths
- Data from other health registries

• Reduce the occurrence of HCM in the breed.

- Encourage HCM scans both before breeding and after 5 years

• Decrease incidence of HD in the breed.

- Encourage breeders to x-ray all their breeding cats before breeding in PawPeds or OFA

• Reduce the risk of entropion.

- Only breed cats with correct anatomy

• Reduce the risk of deafness in white cats

- Solid white cats W- are not recommended to mate to cats with dominant white spotting ws, as the combination may be at higher risk of deafness than Ww.

Long-term goals:

- Define mandatory health tests and requirements for the breed, based on more data.
- Reduce the frequency of HCM in the breed.
  - Ultrasound screening as mandatory requirement in NRR
- Reduce frequency of HD in the breed.

5. Exterior and behavior

5.1 Exterior

As mentioned in Ch. 2 General, the Maine Coon is a breed that traces its origins as a farm cat from the northeastern parts of America. The Maine Coon is therefore referred to as a natural breed where the starting point was a naturally healthy physiology with a certain natural variation in type.

The breed should be large-framed, and with a somewhat square expression in the head and with relatively large ears. The body is distinguished by a wide chest, a solid bone trunk, long and rectangular body ending with a long flowing tail. The breed should be well muscled and have a density that gives your cat a robust and powerful appearance.

It is important that the breed appears harmonious and balanced, and without any distinctive features overshadowing others.

The four major international cat associations FIFe, TICA, CFA and WCF each have their own standards, with a few differences. The standards describe the ideal Maine Coon, and at exhibitions it is the standard the cats are measured against. It is worth mentioning that the different breed standards from the FIFe, TICA and CFA organizations have changed minimally in the last 30-40 years, from the time when the Maine Coon cat was recognized as a separate breed in the various federations that still exist today.

Most Maine Coon breeders in Norway are members of the NRR, and thus it is the FIFe standard that applies first and foremost, and is the one we start from this BBS.

FIFe and the two largest U.S. organizations all have an overarching description in the standard that emphasizes both the origin and appearance of this breed.

FIFe: The Maine Coon is a natural breed of amiable character that traces its origin to the working cat found on the farms of Northeast America.

TICA: The Maine Coon is America's native longhaired cat. The breed, with its essentially amiable disposition, developed through a natural selection process where only the fittest survived. It should always be remembered that the Maine Coon developed basically as a "working cat" able to fend for itself in rough, woody terrain and under extreme climatic conditions.

CFA: originally a working cat, the Maine Coon is solid, rugged, and can endure a harsh climate. A distinctive characteristic is its smooth, shaggy coat. A well-proportioned and balanced appearance with no part of the cat being exaggerated. Quality should never be sacrificed for size. With an essentially amiable disposition, it has adapted to varied environments.

Reference is also made to the breed standards of the other federations for a more detailed description of these.

5.2 FIFe standard

Most breeders in Norway are registered in NRR and FIFe. This BBS is therefore based on the FIFe standard.

The Maine Coon breed was approved in FIFe in 1983, and the standard we have today is essentially the same as that written for the first time; Only minor adjustments have since been made.

General	**Appearance**	The breed of the Maine Coon is large framed with a square outline of the head, large ears, broad chest, solid bone structure, a long, hard muscled, rectangular body and a long flowing tail. Good muscle tone and density give the cat the appearance of power and robustness.
	Size	Large

Head	**Shape**	Medium in size; square outline. Profile with a gentle concave slope.
	Forehead	Gently curved
	Cheeks	Cheekbones high and prominent
	Face/Nose/ Muzzle	Face and nose of medium length with a square outline of the muzzle. Distinct transition can be felt between muzzle and cheekbones.
	Chin	Firm , in vertical alignment with nose and upper lip
Ears	**Shape**	Large, wide at the base. Moderately pointed. Lynx-tufts are desirable. Tufts of hair in the ears extend beyond outer edges of ears.
	Placement	Set high on head with a very slight outward tilt. Ears should be placed one ear's width apart. The width extends slightly in older cats. Lower base set just slightly further back than upper base.
Eyes	**Shape**	Large and widely set. Slightly oval, but not almond shaped, appear round when wide open. Set slightly slanted towards the outer base of the ear.
	Colour	All eye colours, except blue are permitted in any coat colour. Blue is only permitted with EMS codes w, 01, 02 and 03. In any other variety they must be registered as XLH * 61 . Clear eye colour is desirable.
Neck		Males have a very strong muscled neck.
Body	**Structure**	The body should be long, substantial bone structure. Hard muscled, powerful, broad-chested. Large framed, all parts of the body in proportion to create a rectangular appearance.
Legs		Substantial, medium length to form a rectangle with the body.
	Paws	Large, round and well tufted between the toes
Tail		At least as long as the body from shoulderblade to base of tail. Wide at the base tapering to the tip; with full, flowing hair. The hair on the tail is long and always remains flowing.
Coat	**Structure**	All weather coat. Dense. Short on head, shoulders, and legs, becoming gradually longer down the back and sides, with long, full shaggy baggy trousers on the hind legs and belly fur. A frill is expected. Texture silky. Coat has distinct body, falling smoothly. The undercoat is soft and fine, covered by the coarse smooth outercoat.
	Colour	All colour varieties are permitted, including all colour varieties with white; except pointed patterns and chocolate and lilac, cinnamon and fawn. Any amount of white is allowed, i.e. a white blaze, white locket, white chest, white on the belly, white on the paws, etc. For the colour varieties refer to the table below
Condition		The Maine Coon should always be in good balance, condition and proportion.
Remarks		• type must always take preference over colour. • very slow maturing of the breed should be taken into account. • mature males may have larger and broader heads than females. • females are proportionally smaller than males. Allowance must be made for this significant difference in size.

		• length of coat and density of undercoat vary with the seasons.
Faults	**General**	• unbalanced proportions • overall small cat
	Head	• round head • straight or convex profile
	Nose	• nose break
	Muzzle	• pronounced whisker pads • round or pointed muzzle
	Chin	• undershot chin
	Ears	• wide set, flared ears
	Eyes	• slanted, almond shaped eyes
	Body	• fine, light bone structure • short cobby body
	Legs	• long stilty legs
	Tail	• short tail
	Coat	• lack of belly shag • coat of overall even length • lack of any undercoat

5.3 Exaggerated exterior traits

The breed's popularity in recent years also makes the breed prone to exaggerated exterior traits, although the standard still describes a breed that should be balanced and harmonious and without exaggerated traits. Compared to cats 15-20 years ago, you can still see that there have been some changes, especially in the head.

Today we can see a tendency to exaggerate the /angry/feral type, which may have implications for:

- Eyes; more oblique/deep-set. There is also a tendency to see that the eyes have become smaller. It is important to avoid eye anatomy that increases the risk of entropion in the breed.
- Ears; excessive ear-setting – set too high on the head with an ear base that is too narrow.
- Muzzle/chin; Excessive size of muzzle/chin can cause an unbalanced anatomy of the head. The profile line should have a soft curve, and an excessive profile line with marked stop is also an error compared to the standard. A stop in the profile line could also affect the anatomy of the head, where the bones of the skull do not provide ample space for the tear ducts. Excessive focus on chin increases the risk of underbite/other bite defects. A maximum of 2 mm is allowed in FIFe shows, while a correct bite is a scissor bite without malocclusion.
- Size; There is a tendency for excessive focus on size without including focus on health issues that can be exacerbated by the breed getting bigger and heavier. The Maine Coon is one of the world's largest domestic cat breeds, and size is one of the characteristics of the breed in the first place. Typically, a male cat weighs between 6-9 kg, and the female cat somewhat less 4-6 kg. Both smaller and larger individuals exist.

In addition, in recent years in other European countries, new mutation(s) have been bred as well as cross-breeding with other breeds in order to create a new dominant blue eye colour. In 2021, FIFe banned the breeding of blue-eyed Maine Coons, where there is no white or smaller amounts of white (less than 25%), precisely to limit breeding of this mutation(s). At present, it is not possible to test for these new mutations, and we only have the phenotype to deal with. This type of breeding with new/unknown mutations is not widespread in Norway yet, and breeding for new phenotypic mutations is strongly discouraged as there is no overview of the consequences of possible side effects.

- DBE mutation (dominant blue eyes); breeding on an unknown mutation that causes blue eyes, which also apparently produces higher frequency of telecanthus (cf. Waardenburg syndrome). The mutation may also have other unknown health consequences, there is currently little to no research on this mutation and its consequences.

As a counterpoint to exaggerated exterior features, there are also many breeders focused on *preservation breeding*, with the intention of preserving the breed's original characteristics and thus ensuring that today's Maine Coon also has an appearance connection to the original natural breed that it once was. Breeders in Scandinavia are still quite reluctant to include more extreme anatomical features.

5.4 Behaviour

The Maine Coon breed is known to be a breed with a friendly and calm temperament. The cats are sociable towards family members and are often compared to dogs due to the strong attachment to humans. The breed is patient, and often gets along perfectly with children, other cats and dogs. The Maine Coon is considered an intelligent and curious breed, but not infrequently a certain clumsiness is also expressed when exploring new things.

In the breed standards of the Maine Coon, it is often mentioned that the breed is "the gentle giant". A gentle and friendly temperament is a hereditary breed trait, and selecting for good temperament is just as important as selecting for good health and type. The closest we come to "working traits" today is first and foremost being a highly valued pet and family member, who hopefully lives in harmony with us humans in our daily lives.

There are a few studies that have examined cats temperament, behaviour and heredity. They state that temperament is hereditary, and that there is a difference between breeds. Thus, a cat's behavior is determined by genes and experiences that together form its personality (phenotype). Your cat's personality doesn't necessarily influence behavior alone. Environment is also an important factor that controls the motivation behind a behavior.

A 1995 study in England researched kittens' temperament, compared to paternal and maternal temperament, and with socialization based on handling kittens. A kitten has half of its genes from its mother, the other half from its father. In other words, both affect *inheritance*, the genetic, to an equal extent.

A mother who is with her litter and takes care of them as normal also influences the kittens through her follow-up, *environmental influence*.

To measure the *difference* between heredity and environment, one cannot therefore use the mother's influence, since she influences in two ways: both heredity and environment. By looking at the father's temperament towards the offspring, one can measure the degree of inheritance. This does not mean that the father's genes are more important than the mother's genes, but that the father's genetic inheritance can be measured in a study. The mother's genetic influence cannot be studied unless the kittens grow up without their mother.

The study showed that heritage directly reflected how confident the kittens became and how they responded to new objects and situations.

Socialization in combination with genes was most evident in how forward the kitten became with humans and how calm they were when handling. We already know that socialization is important and necessary to have social and loving kittens. When it came to how affectionate a cat gets, the study showed that the combination of socialization and inheritance has equal influence and is equally important.

The parents' genes, shown through the father's temperament, have a major influence on the kitten. Heredity proved to be even more important than environment for the cat to become curious and confident.

Differences between breeds
Both a Japanese study from 2009 and a Norwegian sub-study/master's thesis from 2014 show that there are differences in behavior among breeds.

The characteristics assessed included: activity level, playfulness, sociability towards people, friendliness, curiosity, demands for cuddles, vocalization, aggression towards other cats, aggression towards people, insecurity, nervousness, and defecating outside the toilet box.

In the Japanese study, the cat breeds were divided into 4 groups:

1. High vivacity and high aggression/insecurity
2. High activity and low aggression/uncertainty
3. Very low vivacity and medium aggression/insecurity
4. Low activity and very low aggression/uncertainty

Group 4 that scored lowest on insecurity, nervousness, and aggression towards both other cats and humans, and which also showed low activity levels consisted of the three breeds Ragdoll, British Shorthair and Maine Coon. They scored above average on kindness and demands for cuddles and attention.

5.5 Objectives and strategy

Short-term goals:

• Education in breed history – the past, present and future.

• Breed cats with a good and characteristic temperament.

- The Maine Coon is first and foremost a pet and family member, thus the importance of selecting breeding animals that are confident and social. Aggressive behaviour must be avoided and is not a character trait of the breed.

Long-term goals:

• Change the focus of breeders from "improving" to preserving the phenotype of the breed.

6. Summary

6.1 Important short-term and long-term goals

6.1.1 Overall goal of the breed

It is an overall goal that the Maine Coon cat should continue to be a breed rooted in its origins as the working cat from the farms of Maine, with its robust character and its extremely gentle and good-natured temperament – *the gentle giant*.

6.1.2 Short-term goals

The short-term goals of the breed can be summarized as follows:

- Get more breeders to record all health results publicly
- Decrease the rate of HCM in the breed
- Decreasing the rate of HD in the breed
- Awareness of change in type, avoiding extreme type
- Training, courses for breeders on Breed Specific Breeding Strategy

6.1.3 Long-term goals

The long-term goals of the breed can be summarized as follows:

- Increase the genetic diversity of the breed so that it continues to be as a healthy breed equipped for the future.
- Improving the health of the breed.
- Preserving the phenotype of the breed.
- Certification of breeders

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ATTACHMENT A – Version log

No:	Date modified:	Modified by:	Alteration
1-1	20.04.2023	AMB, TB, BB	Format fix and proofreading
1-2	06-06-2023	AMB, TB, BB, TH	Specifications and improved formulations. Improved goals, removed repetitions. Added sources ch 4.4 and 4.5.