DISTICHIASIS
IN
ICELANDIC SHEEPDOGS

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DIAGRAMS

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**SUMMARY**

Distichiasis is hereditary disorder, with abnormal growth of hair at the eyelids. Tips of these hairs can sting the cornea, resulting in an irritated, becomes inflamed and damaged eye. Distichiasis is found in many dog breeds to a more or lesser extent, and also in the Icelandic Sheepdog (ISD). 47 ISD’s have been diagnosed with distichiasis: 25 dogs in The Netherlands, 11 in Denmark, 7 in Finland, and 2 in both Germany and the United States.

Investigation of the lines of heredity shows, that for 45 dogs at least one ancestor descends from the sire Töfta Njáll (IS 9/81). Njáll produced 48 puppies and 4 of them form the main lines of heredity: Skolli, Sygin, Landi Mestur and Landi Barki.

The lines of heredity at the side of the ancestors of Njáll is non-traceable. What is clear, is that one of his ancestors, Katur frá Keldum (IS 11/68), was used many times for breeding.

His great-grandmother Röskva frá Olafsvöllum (IS 7/73) has offspring with and without distichiasis. The 2 dogs with distichiasis, that do not descend from Njáll, find their origins in Röskva.

Several lines of inheritance are “open”. These open lines have the chance to result in offspring with distichiasis. To what extent the dogs within these lines actually have distichiasis (in case of dominant heredity), or are carrier (in case of recessive heredity) cannot be determined.

The open lines are (fts = frá Thytur Stadir):
1. NL: fts Dimmelimm – Fiskilæðjar Gánska-Noa/Litfari
2. NL: fts Mafur – Van de Wadenborgh Freya
3. NL: Vicky – Anacamptis Dama-Lis/China Joes
4. NL: fts Olafur – frá Fridarstöðum Taktur
5. NL: fts Bersi/Snorri frá Fridarstöðum – Regla frá Isafold
6. DK: fts Katla - Olafur
7. DK: fts Katla - Kormak
8. DK: Oldenhus Lettir – fts Björn
9. DK: Oldenhus Lettir – fts Taco
10. DK: fts Steinunn – Bíbi frá Bjarkarlundi – Surtsey’s
11. SU: Wadsteiner Katla – Tunturiketun Vaskur/Fjella Freki
12. SU: Tunturiketun Asta-Sollilja – Tunturiketun Maggi/Misla/Toa-Freya
13. D: Jarpblesa vom Lechfeld + fts Kormak: Mafur, Mayla, Meyja en Mysla
14. U.S.: Heimskauts Reike – 4 litters with a total of 16 offsprings

Investigation of way of inheritance shows that only genetic factors play a role (Kaufhold et Al, 2009). The exact way of heredity - dominant, recessive or polygenic, could not be confirmed. In mice and humans inheritance is autosomal dominant. When looking at humans, a phenotypic variable expression exists. The cause of the disorder is a mutation of the FOXC2 gene. This mutation in mouse and human results in the “LD – syndrome”, which besides distichiasis also induces lymphedema and varicose veins. When analysing the lines of heredity, more arguments plead towards a dominant rather than a recessive heredity. It seems that, just as in humans, in dogs a phenotypic variable expression exists.

The number of dogs with distichiasis in The Netherlands can be estimated at around 100, so around 4 times more than the actually known 25. In case of recessive inheritance, several hundreds of carriers must be present in The Netherlands.

In The Netherlands breeding with dogs with distichiasis is not allowed. However, dogs with distichiasis are frequently born from distichiasis free parents.

If heredity is dominant, at least one parent must have distichiasis, and if heredity is recessive, both parents are at least carrier.
This implicates, that the outcome of the eye examination is not reliable. In order to test positive, the presence of distichiasis hairs is required. However, in case no distichiasis hairs are found, it doesn't mean that the dog is free of distichiasis. It is possible that this dog is a distichiasis dog without phenotypic expression in case of dominant heredity, and in case of recessive heredity this dog can still be a carrier.

The current breeding Dutch policy (no breeding with dogs with distichiasis) prevents an explosive growth of distichiasis, but is not sufficient to eliminate distichiasis from the population. If we really want to eliminate distichiasis, more measures should be taken. In fact closure of all open lines must be done. In practice this will be difficult. The following policy is conceivable:

1. No breeding with dogs with distichiasis
2. No breeding with the offspring of dogs with distichiasis
3. No breeding with litter mates of dogs with distichiasis

With this measures a small part of the gene pool will be lost. In The Netherlands the factor of inbreeding beyond dogs with distichiasis is rather high, so gene loss will be limited. Sufficient distichiasis free lines will remain, in the Netherlands or alternatively in other countries.

In Denmark an explosion of distichiasis threatens. In Finland, Germany and the U.S. an increase of distichiasis is also possible. To prevent this increase, international agreements should be made, with a minimum of:

1. No breeding with dogs with distichiasis
2. No breeding with the offspring of dogs with distichiasis

Also one should consider not to breed with litter mates of dogs with distichiasis.

Each country should balance these measures against other breeding restraints, based on other genetic disorders. The most severe genetic disorders should – obviously – be given priority.

For a better insight in way distichiasis is inherited, a suggestion would be to re-examine a group of distichiasis free dogs. Think of dogs of the distichiasis lines that are older than 8 years, especially parents and grandparents of offspring with distichiasis.

Finally, genetic research in the form of a pilot study can be considered. The primary goal thereof could be to investigate whether the mutated FOXC2 gene is also present in dogs with distichiasis.
INTRODUCTION

Distichiasis is disorder that involves abnormal growth of hair(s) at the eyelids. These hairs have an ectopic position in relation to the normal hairs on the eyelid, and are growing with their tips towards the surface of the eye (cornea). It may involve only one hair, but a complete row of hairs is also possible. Remarkable is the fact that a location on the upper eyelid occurs two times more often compared to the lower eyelid [7].

These hair tips can sting the cornea, resulting in an eye that becomes irritated, inflamed and even damaged. This inflammation and damage can have a severe impact, resulting in running eyes, frequently blinking, and the eyes may go red. The abnormal hairs are hardly visible without a magnifying glass. The ophthalmologist uses an ophthalmoscope to look at the hairs.

Removing the hairs by depilation serves no use – they will grow back within a couple of weeks. Surgery with excision of the root of the hair by the ophthalmologist is effective.

The prevalence of distichiasis in different breeds varies. There are breeds with a high prevalence, for example the English Cocker Spaniel (37-74%).

Up until this day the cause of distichiasis is not fully understood, but heredity is confirmed. In the English Cocker Spaniel “heredity” (h²) amounts to 0,62 [3]. However, the determination of the kind of inheritance (dominant, recessive or polygene) was not possible in families of Elo dogs [2].

In humans and mice the disorder has a dominant heredity.

Not breeding with dogs with distichiasis is policy in The Netherlands. Nevertheless, dogs that are born from distichiasis free parents may show distichiasis hairs. Is a negative outcome not reliable, and is a false negative result possible?

It is possible that a dog, after having been used for breeding several times with a negative test result, at an older age gets diagnosed with distichiasis (for example Olafur frá Thytur Stadir).

It is also imaginable that a distichiasis positive dog, examined immediately after shedding, shows no distichiasis hairs. And finally, is it possible that a genotypic distichiasis dog never shows the phenotypic expression of this disorder?

In short, distichiasis invokes a couple of questions:

1. Is it possible to distinguish “lines of distichiasis” within the population?
2. Is it possible to get an overview of the kind of heredity?
3. Is it possible to eliminate distichiasis with a breeding policy?
4. Is it possible to formulate an international breeding advice?
5. Is it advisable to organise extra eye examinations for a group of dogs in The Netherlands?
THE ICELANDIC SHEEPDOGS WITH DISTICHIASIS

Within the whole population of ISD’s we find 47 dogs with distichiasis. The Netherlands has the biggest bulk of 25 dogs (see table 1. – left column). In Denmark 11 dogs with distichiasis are known, and Finland has 7. Finally Germany and the United States both have two dogs with distichiasis (see table 2. – left column).

In the Netherlands the eyes of 270 dogs have been examined. About 9% of these dogs has distichiasis. Djarfur frá Thytur Stadir is not mentioned in the table, although a distichiasis hair was found on Djarfurat a certain point of time. However, it was said this hair was caused by a scratch of a cat. In the meantime 17 dogs out of the 25 are older than 10 years, and in the last 5 years only 5 dogs were found positive: each year only one.

I am not familiar with the number of international eye examinations. Sweden examined more than 300 dogs, but not a single dog tested positive for distichiasis. In Norway and Iceland only a couple of dogs have had their eyes examined.

In The Netherlands it is not allowed to breed with dogs with distichiasis. Moreover, the result of the examination is valid for one year only. In Denmark it was forbidden to breed with dogs with distichiasis from 2008 until February 2012. In Germany it is allowed to breed with dogs with this disorder, provided that they are combined with distichiasis free dogs.

### TABLE 1.
DOGS WITH DISTICHIASIS IN THE NETHERLANDS – INBREEDING PERCENTAGE AND LINES OF DESCENT

<table>
<thead>
<tr>
<th>T. = Töfta</th>
<th>O. = Oldenhus</th>
<th>F.O. = frá Olafsvöllum</th>
<th>L. = Landi, other names: frá Thytur Stadir</th>
</tr>
</thead>
<tbody>
<tr>
<td>DOG</td>
<td>%</td>
<td>Sire’s line</td>
<td>Dam’s line</td>
</tr>
<tr>
<td>Anacampsis Enigma Esja</td>
<td>2,0</td>
<td>f.O.Röskva – f.O.Pila</td>
<td>T.Njall – O.Lettir – Katla</td>
</tr>
<tr>
<td>Kaiva’s Askja</td>
<td>1,0</td>
<td>f.O.Röskva – f.O.Prins</td>
<td>f.O.Röskva–Huginn–Djarfur</td>
</tr>
<tr>
<td>Maera</td>
<td>6,1</td>
<td>f.O.Röskva – f.O.Vaskur</td>
<td>T.Njall – O.Lettir – Olafur</td>
</tr>
<tr>
<td>average inbreeding</td>
<td>12,1</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
THE LINES OF DESCENT OF THE DOGS WITH DISTICHIASIS

The question is, are all these dogs with distichiasis related to each other? Is it possible to find similar lines of descent?

In order to look for common ancestors, the origin of all 47 dogs was analysed by looking at both the sire’s side and the dam’s side. Often several lines at one or both sides of the parents were found, but always the choice was made for the shortest line of descent.

First the Dutch dogs were analysed, then the dogs from the other countries. The Swedish dogs were analysed for similarities with the dogs with distichiasis.

<table>
<thead>
<tr>
<th>TABLE 2.</th>
<th>DOGS WITH DISTICHIASIS IN DENMARK, FINLAND, GERMANY &amp; U.S.</th>
<th>INBREEDING PERCENTAGE AND LINES OF DESCENT</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>T.</strong> = Töfta  <strong>O.</strong> = Oldenhus  <strong>f.O.</strong> = frá Olafsvöllum  <strong>L.</strong> = Landi  <strong>TS.</strong> = frá Thytur Stadir</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>DOG</strong></td>
<td><strong>%</strong></td>
<td><strong>Sire’s line</strong></td>
</tr>
<tr>
<td><strong>DENMARK</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fálkastjörnu Sunna</td>
<td>0,2</td>
<td>T.Njall – L.Mestur</td>
</tr>
<tr>
<td>Hi Lydur Ljúfa</td>
<td>1,2</td>
<td>f.O.Röskva – f.O.Vaskur</td>
</tr>
<tr>
<td>Solargeisli Mist</td>
<td>0,4</td>
<td>T.Njall – L.Mestur</td>
</tr>
<tr>
<td>Surtsey’s Askur Árgeisli</td>
<td>2,4</td>
<td>f.O.Röskva – f.O.Prins</td>
</tr>
<tr>
<td>Surtsey’s Örn</td>
<td>5,9</td>
<td>T.Njall – L.Mestur</td>
</tr>
<tr>
<td><strong>average inbreeding</strong></td>
<td>1,9</td>
<td></td>
</tr>
<tr>
<td><strong>FINLAND</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aitiorannan Nordanstjarna</td>
<td>1,2</td>
<td>T.Njall – L.Mestur</td>
</tr>
<tr>
<td>Airiorannan Nypa Nattsoll</td>
<td>1,2</td>
<td>T.Njall – L.Mestur</td>
</tr>
<tr>
<td>Katimoon Mykour</td>
<td>0</td>
<td>T.Njall – L.Barki</td>
</tr>
<tr>
<td>Punaphiljan Visla</td>
<td>0</td>
<td>T.Njall – L.Barki</td>
</tr>
<tr>
<td>Reikikelin Mette-Marit</td>
<td>0,1</td>
<td>f.O.Röskva – f.O.Pila</td>
</tr>
<tr>
<td>Thoka</td>
<td>0,8</td>
<td>T.Doppa – f.Kolsholti Putti</td>
</tr>
<tr>
<td><strong>average inbreeding</strong></td>
<td>1,2</td>
<td></td>
</tr>
<tr>
<td><strong>GERMANY</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>average inbreeding</strong></td>
<td>13,1</td>
<td></td>
</tr>
<tr>
<td><strong>UNITED STATES OF AMERICA</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nor’star Kobbi</td>
<td>2,7</td>
<td>f.O.Röskva – f.O.Prins</td>
</tr>
<tr>
<td><strong>average inbreeding</strong></td>
<td>9,4</td>
<td></td>
</tr>
</tbody>
</table>
**The Netherlands**

In the eighties of the past century, Ans Beer started breeding with two bitches: Frani vom Dennenberg and Cilia vom Hartenberg. They both had the same father: Afram vom Hartenberg. Oldenhus Lettir from Denmark was the male dog that was used for the first litter. Ans also used litter mates of Cilia for the first litters: the bitch Candi and the male dogs Colur and Dagur. It seems logical to start with the analysis of the lines of descent of Afram. You will find them represented in diagram I.

After a lot trying and error, in the end it proved possible to get all lines on one page. However, the result is rather poorly organized. Identifying clear lines is not possible. All dogs are present at least twice in the diagram, while 11 dogs can be found three or more times.

It seemed necessary to look for a more clear overview of the Dutch lines of descent. Since the lines of Afram don't give a clear result, his daughters, Frani and Cilia, are of little use as well. Ans Beer's first litter arrived on april 19 1987. Dam was Cilia vom Hartenberg and sire Oldenhus Lettir. Dogs that were used for breeding from this litter were the male dog Björn and the bitches Katla and Isafold. Of these three dogs, Katla has the most offspring.

Therefore Katla frá Thytur Stadir is choosen for re-analysis.

The lines of descent from Katla are represented in diagram II.

Now, there are only 6 dogs with Katla as ancestor on the both the side of the sire as well as the dam. It concerns: Vicky, Landi Bersi, Chaou van Liangtsou Oska, frá Fridarstöðum Ulka, frá Isafold Tobbi en frá Isafold Bibi. With regards to the other 19 dogs Katla is only represented at the side of the sire or the dam. At 13 of these dogs, we see a descent on the other side of the parents via Oldenhus Lettir and usually via Taco frá Thytur Stadir (8x).

The descent from Katla offers good possibilities to determine lines of descent. A distinction must be made between “open” and “closed” lines. A closed line has no offspring, or it is reasonable that offspring will not occur anymore.

**The closed lines are (FTS = frá Thytur Stadir):**
1. From Krafla to Chaou van Liangtsou Oska
2. From Prydi to FTS Kjartan
3. From FTS Gæfa to Frá Fridarstöðum Veisla
4. From FTS Kiljan to Landi Bersi
5. From FTS Bersi/Snorri frá Fridarstöðum to Fra Isafold Tobbi
6. From Farandi to Vignir
7. From Farandi to Kasper

**There are 7 open lines, with chances of more offspring:**
1. The line of FTS Dimmelimm
2. The line to Frá Olafsfördur Garput-Indasson
3. The line from FTS Mafur to Van de Wadenborgh Freya
4. The line Vicky
5. The line from FTS Ofafur to FTS Fjalladis
6. The line from FTS Ofafur to Frá Fridarstöðum Taktur
7. The line from FTS Bersi/Snorri frá Fridarstöðum to Regla frá Isafold

**1. The line of FTS Dimmelimm**

Dimmelimm 4 descendants that were used for breeding. Two of them (Fiskilækjar Kjartur and FTS Væna) have offspring that most likely will not be bred with. Væna gets Kaiva’s Askja, who tests diastichiasis positive.

The other 2 descendants have offspring with breeding possibilities: **Fiskilækjar Gáskå-Noa** had (together with Indi frá Bjarkarlundi) a litter of 5 in 2010. **Fiskilækjar Litfari** has covered 3 times (2009, 2011) which resulted in 19 puppies (none of them has had an eye examination as of yet).
2. The line to Frá Olafsfloður Garþur Garþur-Indasson

The course of the line is uncertain. There are several possibilities:

- at the side of father’s mother:
  In Denmark Landi Mestur has several descendants with distichiasis, while a Finnish dog also has a line from Landi Mestur to Skovridergaardens Landi

- at the side of mother’s father:
  The line Skovridergaardens Huginn – FTS Djarfur and the line Oldenhus Lettr – FTS Taco are present in several Dutch dogs with distichiasis. Besides this, the distichiasis free status of Djarfur is doubtful.

- at the side of mother’s mother:
  From Farandi (has offspring with distichiasis!) via FTS Gúdrun, Frá Fuglatorgi Grimur and Grimr (all of them distichiasis negative) Grimur had covered 3 times and has 21 descendants: 7 of them have had eye examination and all were negative. Grimr had 3 litters and the only dog that was tested is Garþur-Indasson.

3. The line from FTS Mafur to Van de Wadenborgh Freya

This is a dangerous line. Freya had two litters (2008 en 2011) in combination with FTS Koparker, resulting in 10 descendants. None of this 10 has been involved in breeding until now. FTS Kæpa (daughter of Anacamptis China Joes) was artificially inseminated in the U.S. with semen from Koparker, and got the distichiasis positive dog Heimskauts Reike.

4. The line Vicky

Offspring of Vicky can be divided in 2 lines: Anacamptis China Joes and Anacamptis Dama Lis. Both lines have possibilities to procreate.

AC Dama Lis

Got 1 litter with 2 puppies. One of them, the bitch Anacamptis Enigma Esja, turned out be distichiasis positive after she had a litter. The male dog Anacamptis Gáski, coming from her litter, in the mainwhile has covered two times (a.o. in Germany in 2012).

AC China Joes

This male dog, distichiasis positive, covered three times: once the bitch FTS Tibra and twice FTS Fjalladis (a daughter of the distichiasis positive FTS Olafur).

From the litter with Tibra breeding has taken place with FTS Kaepa, FTS Pjakkur and FTS Prakkari.

FTS Kaepa (bitch)

Went to the U.S., and had 2 litters there (kennel Heimskaut). Meanwhile, one descendant, Heimskaut Reike, turned out to be distichiasis positive.

FTS Pjakkur (male dog)

Covered once in combination with Yra van het Reutse Veld. The litter consisted of 4 puppies.

FTS Prakkari (reu)

Covered 3 times, resulting in 8 descendants (born in 2006, 2008 and 2010).

The 2 litters with Fjalladis resulted in 7 descendants. With 3 bitches (all of them are distichiasis negative) breeding is possible or has already taken place:

- Gáski Frá Ægir:
  Had 3 litters with 10 puppies, not tested (2007, 2009 en 2010).

- Noa Frá Ægir:
  Has 20 descendants (2006, 2009 and 2011); one of them has had an eye examination (distichiasis negative).

- Gæfa Frá Ægir:
  Has 5 descendens (2010), of which Mæra is distichiasis positive.
5. **The line from FTS Olafur to FTS Fjalladis**

In combination with the male dog FTS Djarfur, Fjalladis gets the male dog Frá Ægir Fálki (2002) (distichiasis positive). There has been no breeding with litter mates of Falki. In combination with Anacamptis China Joes, Fjalladis gets 3 bitches, involved in breeding (see also: Vicky – litters from FTS Fjalladis with Anacamptis China Joes).

6. **The line from FTS Olafur to Frá Fridarstödum Taktur**

In the offspring of this line there are 6 descendants with distichiasis. Taktur has 14 descendants, of which Kai Frá Fridarstödum is distichiasis positive.

Breeding has taken place with 3 other descendants from Taktur:
- **Disa-Hófi Frá Fridarstöðum (bitch):**
  - Had a litter with 4 puppies in 2009. One of them, the bitch Glanna Disadóttir Frá Fuglatorgi is distichiasis positive.
- **Snorri Frá Fridarstöðum (male dog):**
  - Covered 3 bitches:
    - Nóa Frá Ægir (2006) – daughter Agnar is distichiasis negative
    - Lýsa Frá Blönduós (2008) – daughter Vínsi Ljósa is distichiasis positive
    - Sumo Frá Isafold (2008) – son Tobbi is distichiasis-positive (the litter of Sumo and Snorri has been closed for breeding)
- **Ró Frá Fridarstöðum (bitch):**
  - Even though she is distichiasis negative herself, she produced in combination with 3 different male dogs 3 distichiasis positive descendants.

7. **The line from FTS Bersi/Snorri frá Fridarstöðum to Regla frá Isafold**

Bibi frá Isafold, a pup of Regla and Snorri, turned out distichiasis positive. Because of this, Regla has been neutered.

**Denmark**

The lines of descent of the Danish dogs with distichiasis are represented in diagram III. It turns out that the lines are starting with descendants from Töfta Njall: Skolli, Landi Mestur and Sigyn. Three dogs only have connection with T. Njall at the side of their mother, while at the side of their father the descent is connected with Róskva frá Olafsvöllum (see also table 2. and diagram IV.)

Most Danish dogs with distichiasis descend from Sigyn – Oldenhus Lettir. With the exception of Surtsey’s Askur Árgeisli, all these Danish dogs with distichiasis have one parent with this descent. Undoubtedly this is related to the many dogs that were imported from The Netherlands, particularly from the kennel of Ans Beer (frá Thy tur Stadir, fts). Imported from this kennel are:
- Brandur, Fokko, Djarfur, Kormak, Olafur, Selfoss, Stigandi and Melrakki.
- Kormak is father of Vicky, a distichiasis positive bitch.
- Selfoss is grandfather of Kormak and covered 8 times.
- Stigandi covered 3 times, and Melrakki 11 times!
- It is possible that Djarfur has distichiasis too. One time during an eye examination one “wrong” hair was discovered, but the story is that this was caused by a scratch of a cat!
- Finally Olafur, who only tested distichiasis positive at the age of 6, covered in Denmark and Finland. His offspring in Denmark and Finland is represented in diagram V.

There are several open lines in Denmark that have the possibility of carrying distichiasis on. It concerns especially offspring from Oldenhus Lettir. Lines are passing fts Björn, fts Taco and from fts Katla to fts Kormak and to fts Olafur.
It is also possible that distichiasis is moving from Germany to Denmark. A descendant from fts Steinunn is Bibi frá Bjarkarlundi and she produced 4 Surtsey litters in Denmark.

As in The Netherlands, we see the same in Denmark: distichiasis is inherited by grandchildren and great-grandchildren. This is particularly clear fts Olafur’s offspring (see diagram V). The question is the status of the dogs between Olafur and his distichiasis positive offspring: are that dogs distichiasis positive or carrier of the mutated gene (depending of the way of inheritance: dominant or recessive)?

**Finland**

The lines of descent of the Finnish dogs with distichiasis are represented in diagram III. Most lines descend from Landi Barki, a son of T. Njall and Mysla. Next to that, there are also lines that originate from The Netherlands through Sigyn – Oldenhus Lettir. From Denmark possible lines are coming in the form of descendants of fts Olafur (see diagram V).

Thoka is not presented in the diagram and is a unique case: It is possible that Thoka descends from Töfta Doppa, sister of Töfta Njall. (line: T.Doppa – frá Kolsholti Putti – Katur). This line is also present in the Dutch Mæra. At the dam’s side the line could descend from Röskva frá Olafsvöllum (via Vaskur frá Olafsvöllum – Kolgrimur). At the sire’s side a remarkable inbreeding is present: Drafn Sorro and Tyra IV are brother and sister (although total inbreeding percentage of Thoka is only 0,8%).

One of the first dogs that was bred in Finland is Tunturiketun Asta-Sollilja (1994). She is a granddaughter of T. Njall, and is present in de lines of 4 Finnish dogs with distichiasis. The offspring of Asta-Sollilja is represented in diagram VII. Only lines of descendants, used for breeding or with the possibility to breed, so born in 2009 or later, are included in the diagram. For this reason the lines of the litters of Asta-Sollilja with the male dogs Ullâlavas Sorti and Hrani are not mentioned.

The 4 descendants with distichiasis are put in a frame. Beside these 4, Asta-Sollilja has another 75 descendants, born in 2009 or later, that have breeding possibilities. So there is a big chance that among these 75 dogs several dogs with distichiasis are present.

It is possible that distichiasis moved from Germany to Finland. A descendant from fts Steinunn is Bibi frá Bjarkarlundi and she produced in Denmark 4 Surtsey litters. One bitch from these litters, Surtsey’s Yra, was used once for breeding in Finland and had 4 puppies Aitiorannan J.J. in 2009.

**Germany**

In Germany there are 2 dogs with distichiasis: Steinunn frá Thytur Stadir and Jarpblesa Vom Lechfeld. Their lines of descent are mentioned in diagram VIII (together with all the other 45 dogs with distichiasis).

Steinunn had two litters. The father of the first litter is A-Vitus frá Himnabær. This litter consisted of 6 puppies. Two of them were used for breeding: Bibi frá Bjarkarlundi (bitch, DCNH IH 112/99, DK 7302/99) and Birthe frá Bjarkarlundi (DCNH IH114/99). Birthe had a litter of 3 puppies frá Bjarkarlundi “G”, but none of them were further used for breeding. Bibi produced 4 Surtsey litters in Denmark. One bitch from this litters, Surtsey’s Yra had 4 pups Aitiorannan J.J. in Finland in 2009. A son of Bibi is Surtsey’s Uggi, who covered 4 times (2008, 2009, 2011).
Another son of Bibi is Surtsey’s Fafnir. He covered 4 times (26 puppies) an has offspring in Denmark (2004, 2005 en 2007) and in Finland (Tunturivetun 2007). A granddaughter of Fafnir, Ishundur’s Hremsa Halla, is distichiasis positive. The second litter of Steinunn consists of 3 puppies frá Bjarkalundi “C”, but none of them were used for further breeding.

Jarpblesa had one litter. She is covered by Kopasker frá Thyur Stadir. There were 4 puppies, born in 2009: Maufur, Mayla, Meyja and Mysla (589/09 – 592/09). Because Kopasker has offspring with distichiasis in the U.S, it is almost certain that these offspring of Jarpblesa has distichiasis too. Several lines of distichiasis of Jarpblesa are possible. At mother’s side they may find their origin in Oldenhus Lettir, while at father’s side the lines may come from Landi Mestur and Oldenhus Lettir both.

It is quite possible that there are more dogs with distichiasis in Germany. Lines of heredity through Oldenhus Lettir may play a role. Lettir produced 2 litters with Candi vom Hartenberg: von Pöttgesberg “A” and “B”. In the past many dogs from The Netherlands were used: from the kennel frá Thyur Stadir (Skotti, Lagsi, Bjalla, Stigandi en Fina), and the kennel Van het Reutse Veld (Astrida, Huginn, Mouk en Mani). All these dogs are offspring from O. Lettir.

United States

In the United States there are 2 dogs with distichiasis: Nor’star Kobbi and Heimskauts Reike. Their lines of descent are mentioned in diagram VIII (together with all the other 45 dogs with distichiasis).

Reike was used for breeding, Kobbi was not. Reike covered 3 bitches and produced 4 litters. The spreading of distichiasis depends of the way of heredity. If it is dominant, theoretically half of the 16 descendants has distichiasis. If distichiasis is recessive, it is possible that the Runestone litter, born in 2011, has puppies with distichiasis. The mother of this litter is Blueridge Mist (DN 07207603). Mist’s grandmother Tryna (IS 2308/91) is a daughter from Vaskur från Olafsvöllum. Vaskur has offspring with and without distichiasis.

Iceland

The start of distichiasis can be traced back to Iceland. All lines come together in Iceland and the dog in the centre is Töfta Njall (see diagram IV.). The parents of Njall, Breki (sire) and Snotra (dam) had 5 puppies. The litter sister of Njall, Doppa, also has two lines of descent (to Thoka in Finland and to Maera in The Netherlands).

Analysing the ancestors of Njall to detect the origin of distichiasis is not possible. Often Katur frá Keldum was used to litters, and the degree of inbreeding of Breki is very high (37.5%). In combination with Tyra, Njall produces Skolli and Sigyn, and by covering Mysla, Landi Mestur and Landi Barki come to existence. Almost all Dutch dogs with distichiasis are descending from Sigyn, while the Finnish dogs mainly descend from Landi Barki. The Danish dogs have a “spread source” with descendants from Skolli, Sygin and Landi Mestur. The Swedish Icelandic Sheepdogs, all distichiasis free, descend from Garda Grettir. Grettir is only indirectly related to Njall.
Töfta Njall produced a total of 12 litters (see diagram IX). The litters with offspring with distichiasis are underlined. All grandchildren of Njall are mentioned. It is nice to see that many Danish dogs have a distichiasis free line of heritage. The Netherlands get the most offspring with distichiasis from Oldenhus Lettir, while the line from Landi Grana to frá Husavik remains distichiasis free. Finland started breeding ISD’s around and about 1992, and the distichiasis line there originates in Landi Barki, although many dogs got imported from Denmark also.

**Sweden**

Sweden examined the eyes of more than 300 dogs, but distichiasis was found. Analysing the origins of the Swedish dogs is therefore interesting. A clear overview can most likely be produced when looking closer at the dogs that had their eye examinations in the first years these were taken. A random check is necessary and consists of 15 dogs, born in 1999 or previous years, where possible from different kennels. It concerns a total of 137 dogs with eye examinations, born in that period. The 15 randomly chosen dogs, all from different kennels, are represented in table 3. Striking is the fact, that there is not one dog that descends from Töfta Njall. Most dogs have a line of descent from Röskva frá Olafsvöllum, either via Vaskur frá Olafsvöllum to Garda Grettir (see diagram IV.), or via Lappi frá Olafsvöllum (IS 66/79). Lappi is a son of Röskva and Kolur frá Olafsvöllum (Sætúni, IS 12/68). Remarkable also is that Snær frá Keldnakoti (IS 44/80) is present in several dogs as ancestor. Their are two dogs (Jara and Ulvdalens Jaki ) without a line of descent from Röskva at mother’s side, but at that side Snær is found in that line.

**Norway**

I have no information about dogs with distichiasis in Norway. When analysing the database, I didn’t find any Dutch dogs that were used for breeding in Norway. The dogs in Norway find their origin in 4 Icelandic dogs: Snær frá Keldnakoti (IS 44/80), Pila frá Olafsvöllum (IS 10/76), Lappi frá Olafsvöllum (IS 66/79) and Garda Grettir (IS 22/81). Pila and Lappi are brother and sister coming from Röskva (IS 7/73) and Kolur frá Olafsvöllum (Sætúni, IS 12/68). The lines in Norway are comparable with the ones of Sweden. It seems quite likely that Norway has no dogs with distichiasis.

**Lines of descent and Inbreeding**

There are striking differences between the countries with regards to the extent of inbreeding. The average inbreeding percentage of dogs with distichiasis is represented in the tables 1, 2 and 3. The low degree of inbreeding in Finland (1,2%) and Denmark (1,9%) may have something to do with the fact that the dogs have a large number of lines of descent from T. Njall, while the Dutch dogs are mainly descend from Sygin. In Sweden (13,0%) 12 of the 15 distichiasis free dogs are descending from Garda Grettir (diagram IV).
TABLE 3.
RANDOM SAMPLE OF 15 SWEDISH DISTICHIASIS NEGATIVE DOGS – INBREEDING PERCENTAGE AND LINES OF DESCENT

\( fO = f\)rá Olafsvöllum  T = Tófta  G = Garda  S = Skovridergaardens
Röskva = frá Olafsvöllum Röskva  Tinni = Garda Tinni  Loa Yr = Garda Loa Yr
*) unable to determine descent from T.Njall, T.Doppa or f.O.Röskva

<table>
<thead>
<tr>
<th>DOG</th>
<th>%</th>
<th>Sire’s line</th>
<th>Dam’s line</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boklättens Bordi</td>
<td>8.2</td>
<td>Röskva – f.O.Vaskur – G.Grettir</td>
<td>Röskva – f.O.Pila</td>
</tr>
<tr>
<td>Jara</td>
<td>28.6</td>
<td>Röskva – f.O.Lappi</td>
<td>---- *)</td>
</tr>
<tr>
<td>Ulvdalens Jaki</td>
<td>16.4</td>
<td>Röskva – f.O.Lappi</td>
<td>---- *)</td>
</tr>
<tr>
<td><em>average inbreeding</em></td>
<td>13.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>frequency of the lines</td>
<td></td>
<td>Röskva – f.O.Vaskur – G.Grettir</td>
<td>13</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Röskva – f.O.Lappi</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Röskva – f.O.Vaskur – T.Njall (jr)</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Röskva – f.O.Hrafnafloki – S.Kolur</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Röskva – f.O.Kubbur</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Röskva – f.O.Vaskur – Loa Yr</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Röskva – f.O.Pila</td>
<td>1</td>
</tr>
</tbody>
</table>
HEREDITY – DOMINANT OR RECESSIVE?

Kaufhold et al. analysed the way of heredity of distichiasis in ELO dogs. It concerned 7 families, with 218 examined dogs, where 65 (30%) were diagnosed with distichiasis. A model of inheritance was set up, but the way of heredity (dominant, recessive or polygene) could not be determined. A model only including non-genetic factors could be excluded [7].

In mice and humans we see an autosomal dominant heredity. The cause is a mutation of the FOXC2 gene [5,6,8,13].

In humans this mutation is responsible for the so-called “LD-syndrome” (lymphedema-distichiasis syndrome) [1]. This syndrome has a variable expression, and a variable age of disclosure. It often starts during puberty [1,4,8,13]. It is possible that distichiasis manifests itself alone, but usually the symptoms of lymphedema are presenting themselves later on as well. In about half of humans with distichiasis the lymphedema expresses itself in venous insufficiency with varicosis [9,13]. The valves of the veins in the legs are malfunctioning. In mice, hyperplasia of the lymph vessels is found, but seldom hind limb swelling is seen [8].

Additional symptoms of the LD-syndrome are possible at the eye: conjunctival edema, photophobia, exotropia, ptosis, congenital ectropion and congenital cataract [4,10]. Other complications are: cardiac defects, cleft palate and extradural cysts [4].

In cats, distichiasis is considered rare. A 2 years old cat suffered from bilateral eye discharge and blepharospasm. At examination conjunctivitis, ulcerative keratitis and distichiae were found. Several treatments didn’t work. After more than one year the distichiae were surgically removed, resulting in complete resolution of the clinical signs [12].

In dogs, the way of heredity is not clear. An argument pleading for recessive heredity is the fact that in all 47 dogs with distichiasis at both sides of the parents a line of descent can be found. Dogs, positioned on those lines would at least be carrier of the mutated gene.

A recessive heredity has a lot of consequences.

In The Netherlands about 270 dogs have had eye examinations, and 25 (9%) were diagnosed with distichiasis. At this moment in time the total amount of dogs registered in The Netherlands is about 1400. At the time the first eye examinations were taken, the number was around 1100. In other words: 270 of the 1100 dogs have been examined. This is about one quarter. This means that in The Netherlands the total amount of dogs with distichiasis would be 4 x 25 = 100.

With recessive heredity, both parents are at least carrier, and half of offspring is carrier as well. The average size of a litter is 5,5. So the amount of carriers in The Netherlands is: 2 + 2,75 = 4,75 x 100 = 475!!

We can therefore state that in case of recessive heredity, the amount of carriers in The Netherlands is at least a couple of hundred.

There are more arguments which plead for dominant heredity:

1. In mice and humans heredity is also dominant.
2. The analysis of lines of descent shows that often the line at one side of a parent is short, while the line at the side of the other parent is much longer.
3. In humans the dominant LD-syndrome expresses itself often at later age. The same can be seen in ISD: although initially no distichiasis is found, later on distichiasis hairs may manifest themselves.
4. Tófta Njall has had 12 litters with a total of 48 descendants. Twelve of them were used for breeding. Only 4 of these 12 passed distichiasis on through their offspring, the remaining 8 didn’t (see diagram IX).
Now, there are 2 possibilities:

1. **in case of recessive heredity:**
   - The 4 descendants (Landi Barki, Landi Mestur, Skolli and Sygin) are carrier and the other 8 are not. In this case dogs with distichiasis should always have two lines of descent to 2 of the 4 carriers, or:
   - The 4 descendants have distichiasis and the other 8 are carrier. In this case there should be a couple of dogs be found that descend from 2 of the 8 carriers.

2. **in case of dominant heredity:**
   - The 4 descendants (Landi Barki, Landi Mestur, Skolli and Sygin) have distichiasis and the other 8 have not. This is the most self-evident situation.

The survey of lines of the Dutch dogs with distichiasis is adapted to the situation of dominant heredity (diagram X). The most logical (shortest) lines are chosen, and Oldenhus Lettir, son of Sygin, is taken as starting point.

A problem exists at Garpur-Indasson frá Olafsfjörður. Several lines of descent are possible. Besides Fina frá Thyttur Stadir, who is mentioned in the diagram, other possibilities are:

- Landi Mestur – Skovridergaardens Landi – Agnea and Indi frá Bjarkalundi
- Farandi – Gúdrun frá Thyttur Stadir – Grimur and Grima frá Fuglatorgi
- Oldenhus Lettir – Taco and Trissa frá Thyttur Stadir – Skjalla frá Gull Lyklinum

The survey (diagram X) shows 25 dogs with distichiasis, and 14 dogs without distichiasis. Is it true that these 14 dogs don’t have distichiasis, or do they have only the genes for distichiasis, but don’t have the phenotypic expression?

**Conclusion**

Although there are more arguments for a dominant heredity, it cannot be proved. However, it is plausible that the ISD has a dominant heredity for distichiasis with an incomplete phenotypic expression. Expression may start at later age, or never!
BREEDING POLICY

In The Netherlands the actual policy is not to breed with dogs with distichiasis, but frequently dogs with distichiasis are born from distichiasis free parents: Bibi and Tobbi fra Isafold, Garpur Indasson fra Olafsjödur and Maera. In case of dominant heredity one parent must have distichiasis, and in case of recessive heredity both parents must be carrier. Obviously the Dutch policy is not able to eliminate distichiasis, but only it prevent expansion (in relation to the increase of the total population).

The example of the distichiasis offspring of distichiasis free parents implicates, that the result “negative” of the eye examination is not reliable. Only “positive” is a reliable outcome.

Is it possible to eliminate distichiasis through a breeding policy out of the ISD population?

If elimination of distichiasis is desired, all open lines of descent should be stop breeding. The following measures also need to be taken:
1. No breeding with dogs with distichiasis
2. No breeding with offspring of dogs with distichiasis
3. No breeding with litter mates of dogs with distichiasis

These measures seem to be sufficient. If in the future more measures have to be taken, breeding with offspring from grandparents with distichiasis positive litter mates should also be stopped.
With the three abovementioned measures a small part of the gene pole will be lost. For The Netherlands the lost will be minimal because of the high inbreeding percentage of dogs with distichiasis – the dogs are too closely related anyway. In The Netherlands as well as in other countries enough distichiasis free lines remain.

Is it possible to formulate an international breeding policy?

In Denmark an explosion of dogs with distichiasis threatens. Without a change of strategy, we will see also an increase of dogs with distichiasis in Finland, Germany and the United States.
If one want to stop this increasing, it is necessary to:
3. Stop breeding with dogs with distichiasis
4. Stop breeding with offspring of dogs with distichiasis

Perhaps with these measures it’s possible to stop distichiasis in Finland, Germany and the United States. For the situation in Denmark and The Netherlands this policy as its best can stop the spreading, but distichiasis will remain in these countries. Perhaps it is possible to come to an agreement regarding a communal policy within I.S.I.C. However, it is important to consider the impact of distichiasis in relation to other, perhaps more serious, genetic disorders, in taking discisions about what must be done.

Is it advisable to examine more dogs?

Getting more insight in heredity is possible if a sample group of older dogs, in the past distichiasis negative, will undergo a re-examination of the eyes. Think about dogs that are 8 years or older, parents or grandparents from dogs with distichiasis.
We must also consider the option of genetic research. A pilot study is possible to analyse if the FOXC2 gene, responsible in humans for distichiasis, also plays a role in dogs.
LITERATURE


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