

Recommendation for breeders

How to deal with POAG/PLL in breeding blood lines

Glaucoma and Primary Lens Luxation are serious eye diseases that can cause vision loss...

Written by Valerie Lore

You have probably heard about POAG/PLL in Shar-Pei.

Primary Open Angle Glaucoma (POAG) and Primary Lens Luxation (PLL).

Both are considered as autosomal recessive diseases and appear to be caused by the same mutation. The guilty gene is correctly identified (2017) and named ADAMTS17 gene.

The diseases were not well known in the last two decades, but now, both became one of the main concerns for Shar-Pei communities.

We don't know exactly the origin of this mutation and how this disease appeared in our breed, but in nowadays lots of new cases of eye problems are detected by ophthalmologist veterinarians around the world, reported to be relative to POAG /PLL disease. These serious health problems should be highlighted for a better understanding and learning, how to handle it in our breeding programs.



AHT pictures

What is POAG? What is PLL?

We know the disease occurs in very early age, reported to start between 7 and 8 months by unsightly and painful changes visible on an eye ball.

POAG (primary open angle glaucoma) is characterized by reduced drainage of fluid from the eye. This causes a build-up of intraocular pressure which, in turn, can lead to pain and blindness. It is possible to observe enlarged pupils, cornea swelling, and scleral congestion.

The eye widens with abnormal volume and sometimes can have different sizes due to pressure.

PLL (primary lens luxation) is characterized by lens dislocation and can also lead to loss of vision.

MAIN SIGNS THAT SHOULD ALERT YOU

If your Shar-Pei shows one or several symptoms described below, or if your Shar-Pei starts to have irritated eyes and shows discomfort, be vigilant.

Do not think, that it can be just eyelash friction, the beginning of keratitis, or better the beginning of entropion.

Whatever it is, you should visit the vet for a professional examination to define the right diagnosis.

Do it as soon as possible. **all eye problems should be treated as serious.**

Warning symptom:

- Rubs eye
- Shows discomfort
- Apparently poor vision
- Teary eyes
- Sometimes difficulties with recognizing friends
- Easily frightening can indicate problems with lens luxation



“ONLY a precise ophthalmological examination can define real eye problems and make diagnosis.”

Diagnosis and confirmation of POAG

To diagnose the dog's glaucoma, the veterinarian examines the eyes and controls the eye pressure by using a tonometer. A closer examination of the inside of the eye can be done by using specialized instruments such as an ophthalmoscope or a gonioscope.

- **Tono-pen/Tono-vet:** Measures intraocular pressure
- **Examination** of the fundus as far as possible
- **Gonioscopy:** Evaluates the irido-corneal angle of the contralateral eye

The function of the retina is also evaluated by an ophthalmologist using the electroretinogram.

Long-life treatment

A Long-life treatment aims to keep vision as long as possible.

The medical treatment of glaucoma consists of different kinds of ophthalmic drops. First of all, it is important to reduce pressure in the eyes, but also to treat pain.

However, despite topical treatment to lower intraocular pressure, it may continue to increase.

Vision still functional, Primary Glaucoma

In case of intraocular pressure moderately controlled with medical treatment or in prevention of primary glaucoma that has already affected the other eye, surgical treatment to keep vision available.

These procedures can be combined or carried out individually:

- **Transscleral cyclophotocoagulation** with diode laser: Decrease in intraocular fluid production (aqueous mood)
- **Ahmed implant**: Unidirectional valve ensuring control of intraocular pressure (< 7-8 mmHg)

Absent vision - Primary Glaucoma

If vision is absent, medical treatment does not control intraocular pressure and your pet feels pain and discomfort. In this stage enucleation (full withdrawal of the eye) or evisceration (removal of the contents of the eye and placement of an intrascleral implant) are recommended.

It is possible to have a fluctuation of intraocular pressure during the day. Chronic pain is often present in these cases.

Knowledge of clinical signs is important in order to address this condition as soon as possible and thus will limit the loss of visual function.

The consequences of glaucoma in dogs

This disease is much more complicated in dogs than in humans and can cause blindness to the affected eye.

In cases where it is a hereditary disease, the second eye is also affected, which is why it is treated with prevention. If the pressure cannot be reduced with medication, it is better to perform eye surgery of the dog with surgical removal of the eye to prevent the animal from suffering.

A genetic inherited disease

POAG/PLL is an inherited genetic disease. Each parent is, or may not carry the gene of this disease. Carrier can transmit copies of gene to the offspring.

Examples:

If a male **clear for POAG/PLL** (not carrying the gene of disease), will be mated to a female, **carrying** a copy of the gene, then they can produce puppies that will be **clear** for the POAG/PLL

and also puppies that will carry one copy for POAG/PLL. Puppies with one copy will be called **CARRIERS**.

This kind of mating is possible without the risk to get affected puppies in the litter.

On the other hand, if the male and female are both carriers (they each have a copy of the gene) they could have clear puppies, carrier puppies, but the worse scenario of this mating is production of affected puppies in the litter. **It is reason, why this combination of parents is not recommended.**

!! Before using males and females in the breeding program, we should know important information about the number of copies which they carry / or not...!!

⊗ STOP THE SPREAD IN THE BREED. ⊗

How to do it?

The only reliable way to know if a dog IS or IS NOT a carrier of the gene is TESTING!

There is a DNA test called POAG/PLL. By salivary (or blood) sampling we can reliably detect dogs that are carriers, affected or free of the problematic genes.

Breeders can choose parents for future puppies taking into account both results for POAG/PLL to avoid that affected puppies will be born. This test is the only way to decrease the number of affected dogs.

Before starting all breeding projects, testing both (the male and the female) is highly recommended and mandatory for responsible breeders.

Future parents must be compatible.

Where find DNA tests?

In Europe POAG/PLL tests are offered by several laboratories.

The mostly used laboratory is Laboklin. You can order the swab kit for POAG/PLL on the website from each laboratory offering the tests.

You will receive your test kits with the application form to fill in all data that the application form is asked for. The data should be fill in by your veterinarian but you can also do it yourself.

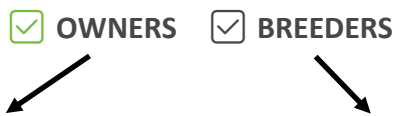
The swab sample should be done by your veterinarian. It is fast and absolutely not painful.

You have to send two samples + the document to the laboratory.

You will receive a result by email or by letter approx. in 1 week.

WHO SHOULD TESTING?

RESPONSIBLE PERSON TEST BEFORE THEY START WITH BREEDING



Are the major actors in the early detection of this serious diseases.

Testing our breed stock is an effective method for detecting problems and taking wise decisions in our breeding program.

——> TEST CAN BE ALSO DONE BY PET OWNERS IF SOME DOUBTS OR SYMPTOMS ARE ALREADY PRESENTED.

"Better safe than sorry"

Understand results from POAG/PLL test How to handle with results in breeding?

The appearance of the POAG/PLL in Shar-Pei has significantly changed the way we plan our projects. Our vision of the healthy dog has changed too.

Before, it was rather simple to find the right partners for the long-awaited match by bringing all sought criteria such as longevity, health, beauty, and character with total respect of standard....

In the past, the knowledge of pedigrees was the best way and main source of information that we have used to make the right breeding decisions to improve or correct breeding programs.

However, knowledge to read pedigrees allows us to know valuable insights that we can use for calculating inbreeding, taking notes about colors, coats and so many more

Currently, lots of responsible breeders and owners feel concerned about the health problems that our breed is going through.

Lots of them already tested their dogs for SPAID, HIPS, ELBOW, COLOR, BEAR COAT, THYROID and many more never forget that we have a common interest to test as much as possible.

For now, we still follow the right way to reach the expected goal: Trying to Produce nice and healthy Shar-Pei, even though the POAG /PLL is complicating our mission.

Actually, many stunning dogs could be compatible but they are not or are no longer because of the transmission of this gene.

Selective breeding must respect the results of science but not by removing all carrier dogs away from breeding programs.

This would be highly damageable to the breed existence and its diversity. We will lose a large part of the existing gene pool.

Step by step, we will see decreasing in the mentioned eye problems. Thank DNA TESTS we can do wise decisions to breed or not.

Let's do it all together and let's hope this nightmare will be over in some years.

Test in the early stage of puppy life is the best solution and our advantage.

UNDERSTAND RESULTS FROM POAG TEST

**N/N RESULT
=
CLEAR FOR POAG**

Your dog has not inherited any copies of the gene, so he is free of the disease and cannot transmit it. He is therefore **NOT CARRIED**

The readable results will be

N/N

Or NON-CARRIER / CLEAR

HOMOZIGOT

IT CAN BE MATED

✓ with dogs carrying the gene

✓ With non-carrier dogs like him

✓ With affected dogs

**N/POAG RESULT
=
CARRIER**

Your dog has inherited a copy of his parents' gene, he will transmit a copy of the gene to his offsprings, he's called a **HEALTHY CARRIER**.

The readable results will then be

**N/ POAG or CARRIER /
HETEROZYGOTE**

IT CAN BE MATED:

✓ with dogs **NOT CARRIER** (free)
result **N//N - CLEAR**

But **NEVER WITH DOG WITH
SAME RESULT AS HIM N/POAG OR
POAG/POAG**

**POAG/POAG RESULT
=
AFFECTED**

Your dog has inherited 2 copies of his parents' gene, he will develop the disease and transmit 2 copies to his offsprings, the dog is then **AFFECTED** by the POAG

Readable results will be

POAG/POAG or AFFECTED

IT CAN BE MATED

✓ with dogs **NOT CARRIER** (free)
result **N//N - CLEAR**

● Or removed from breeding program if the selection criteria are not met

✓ The tables below can help to avoid misunderstandings

Keep in mind!

✓ Most DNA-tested dogs can be used responsibly in a breeding program, but the decisions you make when choosing dogs to mate them must be carefully planned.

✓ Choices and decisions are personal and engage only those who make them. Think twice, and make a risk assessment before you start to act.

✓ Don't remove from breeding programs all carriers dogs. Decreasing the number of affected and carrier dogs can be done with time. The best approach is to put DNA tests into breeding practice and learn how to use the results.

Note: Testing a full litter certainly means spending also but allows you to select your future keeper very fast considering the best results, and allow you to reduce the spread of POAG/PLL in your blood line.

Below is an explanation of heredity POAG and PLL and up to date statistic data from Laboklin Germany. The data shows how POAG and PLL are spread across the population of SHAR- PEI in Europe.

TAKE INTO ACCOUNT RESULTS FOR BREEDING PROGRAMS

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	MALE CLEAR FOR POAG N/N	CARRIER MALE N/ POAG	AFFECTED MALE POAG POAG
CLEAR FEMALE N/N	ALL PUPPIES WILL BE CLEAR N/N	50% CLEAR PUPPIES N/N 50% CARRIER PUPPIES N/POAG	ALL PUPPIES WILL BE CARRIERS N/POAG
CARRIER FEMALE N /POAG	50% PUPPIES N/N 50% PUPPIES N/POAG	25% PUPPIES N/N 25% PUPPIES POAG/POAG 50% CARRIER PUPPIES N/POAG	50% CARRIER PUPPIES N/POAG 50% POAG/POAG AFFECTED PUPPIES
AFFECTED FEMALE POAG/ POAG	ALL PUPPIES WILL BE CARRIER N/POAG	50% CARRIER PUPPIES N/POAG 50% POAG/POAG AFFECTED PUPPIES	ALL PUPPIES WILL BE AFFECTED POAG /POAG

Very important note: In case of an accidental mating between two carriers or affected dogs, it is very important to test all puppies before they will leave for their new homes. Veterinary advice will be important to ensure appropriate care for their health and good quality of life.



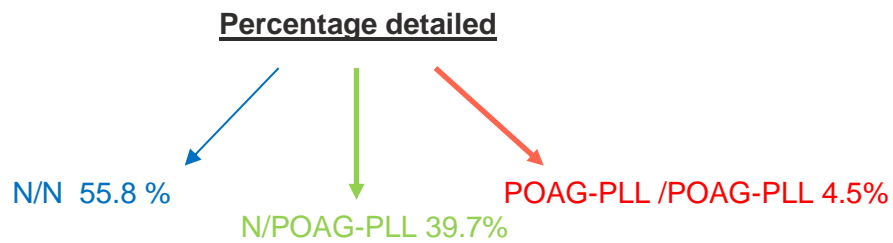
**Statistics of where we are
and how many dogs we have tested**



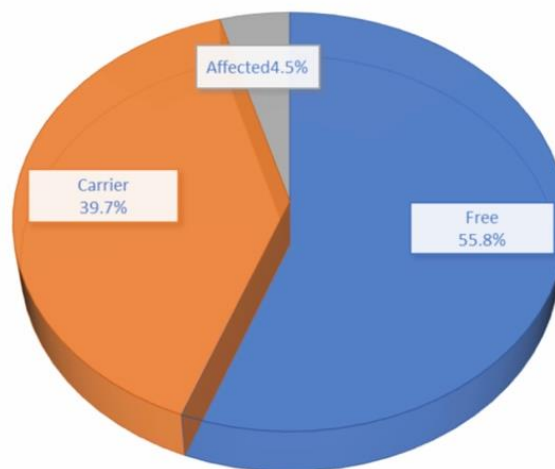
Based on samples from European countries in LABOKLIN, the official report presented below shows the number of Shar-Pei tested for POAG/ PLL and the proportionality of the results: Clear, Carrier and Affected.

Statistics up to May 2022

1300 Shar-Pei tested



POAG/PLL UP TO 5_22 EUROPE_LABOKLIN



** unconditional breeding consideration / unconditional development of a breeding strategy at a allele frequency > 20 % (allele frequencies < = 5-10 % are desirable)*

** notes marked with * were added by the author. The notes base on telephone information from Laboklin on demand for correspondence. 08.22*

Link to the full text of the report:

https://www.efspc.eu/wp-content/uploads/2022/09/SEC_Final_EFSPCSPAID-EN.pdf

References:

1. James A. C. Oliver, Sophie Rustidge, Louise Pettitt, Christopher A. Jenkins, Fabiana H. G. Farias, Elisabeth A. Giuliano, Cathryn S. Mellersh (2018) A novel ADAMTS17 mutation is associated with primary open angle glaucoma and primary lens luxation in the Shar Pei.
2. CHUV DE Montréal By Dr. Maria Vanore, Dipl. ECVO, clinician-teacher in ophthalmology anicura.fr CHV regis.com / DR. PAYEN, Ophthalmology Specialist
3. ENVA-National Veterinary School Maison Alfort/ ophthalmologist service