

Polymyositis

Basics for the Kooikerhondje Community



A Health Concern In Breeding Lines

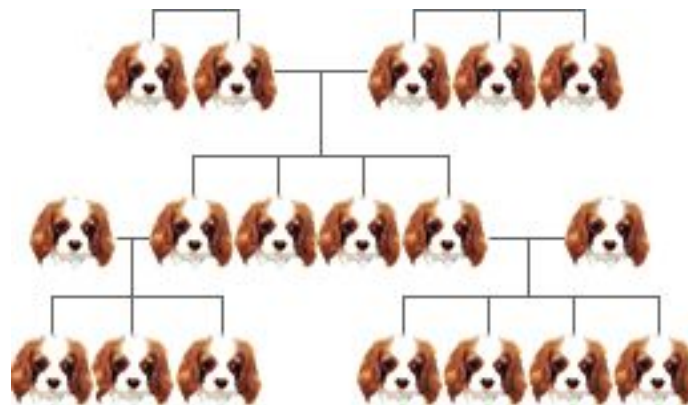
Research summary and case report

By Susanne Martin, MD

The word “Polymyositis” has been floating around in the Kooikerhondje community for many years now. It is a rare but scary disease and can be lethal in months to years. There is no test available and it is not known if this disease is familial and/or hereditary in the Kooikerhondje.

Polymyositis has been diagnosed more often in recent years. This article is a summary from multiple publications and presentations of Prof. Mandigers and his team of the University of Utrecht in the Netherlands. Their research to understand the nature of this illness and to hopefully find a genetic marker continues and donations are appreciated (see info at the end of this article).

Heritable Diseases in the Kooikerhondje:



- **Hereditary Necrotizing Myelopathy (HNM) or also called ENM - DNA test for carrier status available**
- **Von Willebrand's disease (vWD) - DNA test for carrier status available**
- **Polymyositis (PM) - no test available - pedigree/registry research and careful use of lines/dogs that have produced the disease**
- **Epilepsy (EP) - no test available - pedigree research necessary**

These diseases have an effect of the effective population size and average increase of inbreeding per generation.

FACTS:

The Kooiker dog is an old Dutch breed originally used to catch ducks in a decoy. After the Second World War, 42 of these dogs remained of which only 14 animals, and especially **10 of them, were used to re-establish the breed**. The foundation is herewith small.

Polymyositis, short PM (>60 cases diagnosed and reported in 2016) causes **chronic muscle inflammation** of one or more muscle groups resulting in loss of functionality and muscle weakness in the affected areas. PM is progressive, has no gender disposition, and has a very poor prognosis. Some affected Kooikers are relatively young and seem to present with more swallowing and eating related problems. Another group is middle aged and has more musculoskeletal problems and sometimes a combination of both.

Most common symptoms:

Reduced endurance, muscle weakness, fatigue, lack of drive to play or walk, lameness, stiffness, swallowing problems including salivation, vomiting, poor appetite, and walking with a curved back.

Abnormal markers in the blood include **elevated CPK** levels (an enzyme called creatine phosphokinase which is important for muscle function), **elevated liver enzymes AST/ALT and elevated protein in the urine** besides others.

Because many symptoms are also features of other disorders, this condition is often **difficult to recognize** and can be easily misdiagnosed.

Besides potential genetic/autoimmune origin for PM, infections and toxins can also trigger general myositis that doesn't present as progressive and generalized.

In order to make a **definitive diagnosis, a muscle biopsy** by a DVM (general or subspecialized) is required. Other medical work-ups include an electro-myography (test to measure the muscle reaction after a stimulus is given) and bronchoscopy (internal exam with a camera visualizing the lungs and bronchial tree if there is a difficulty swallowing).

If diagnosed early, the disease progression can be slowed down or go into remission. Available treatments are immunosuppression including management with steroids. Complementary therapy and supplements can bring some relief as well.



CASE: Maggie (4/2011 - 7/2016)

Maggie was born in the Netherlands at a known kennel and moved to the USA at around 8 weeks of age.

She was an easy going and sweet girl. She liked other dogs and never really caused trouble. She passed all of her 5 health clearances and received a CHIC number. She had two litters - the first one with 4 puppies and the second one with 5 puppies. She was a very good mother. All puppies are happy and healthy to date.

5 months after her second litter, Maggie seemed less interested in participating in training classes. She hesitated to jump into the hatch back of the vehicle. She seemed more lethargic and watched the other dog play in the garden while laying in one favorite spot. Something seemed truly wrong when Maggie didn't enjoy her usual hike and she kept the base of her tail tucked close to her body. She was brought to the vet to rule out a urinary infection. Many blood panels were obtained including for tick-borne illnesses. All came back negative. Maggie's energy level went down further over the next few days. She was brought to a different veterinarian. Given her high level of activity prior to onset of symptoms, a CPK level was obtained to look at muscle breakdown and the number was very high at 4544 IU/L (normal: < 895), (AST 141 IU/L with normal < 66, ALT 139 IU/L with normal < 118) and protein 2+ in the urine. Luckily, the owner sought advice from seasoned Kooikerhondje owners with experience in the various Kooikerhondje related health

issues. . Due to the elevated CPK level, the concern of underlying Polymyositis was raised immediately. The owner was advised to get a muscle biopsy done. The **biopsy confirmed the disease in July 2015** (head, shoulder, lumbar and rump samples: mixed-inflammatory infiltrate and varying degrees of myofiber degeneration, necrosis, and regeneration; morphological diagnosis: lymphohistiocytic and neutrophilic myositis), one month after her first symptoms. At that point, **Prof. Mandigers at the University of Utrecht in the Netherlands got involved remotely to help guide** Maggie's further care. Maggie was started on **oral prednisone (a steroid)** to help her symptomatically by suppressing the degree of inflammation.

Blood work to monitor the CPK level, kidney function and liver enzymes was obtained every 4 weeks.

Over the following months, the owners tried many different **complementary therapies** including weekly swimming, massages (with Jin Shin Jyutsu showing the best result in terms of mobility), laser-acupuncture (to reduce inflammation and pain). **Complementary medicine** included **supplements**: vitamins, milk thistle, salmon oil and others and **herb blends**: Chinese herbs but also cinnamon and ginger.

Raw food was not recommended due to her compromised immune system and the owners cooked for the months after diagnosis a grain free diet with low glycemic vegetables mixed in with meat.

Despite all efforts, Maggie **continued to get steadily worse**.

Maggie enjoyed participating in daily life by going on rides in her **wagon** when walking became more difficult.

Several months into the progression of the disease, her digestive system slowed down and was more affected. She became **incontinent** both for urination as well as bowel movements.

In the last month of her life, she was started on Flagyl (Metronidazole). It is an antibiotic that helps with diarrhea and helped with Maggie's symptoms.

Maggie was put to sleep in July 2016 when her quality of life decreased and her discomfort worsened. This was one year after her diagnosis of PM. She was cared and loved for to the very last moment. Her family went through so much sorrow but did everything they possibly could have done.

Many cases of this terrible illness are never diagnosed or remain hidden to the public.

We wish to raise awareness to all owners and breeders. **Dogs with symptoms listed below need further work-up and should be reported to the VHNK Registry.**

The current research and work of Prof. Mandigers is the only place we are hoping to find an answer about inheritance of this disease and hopes for a genetic test are

high.

If you are interested in supporting his research, please don't hesitate and donate to the account at the end of this article.

IN A NUTSHELL:

SYMPTOMS

And what should raise your attention

- Reduced endurance, general fatigue
- Lack of drive to play or walk
- Muscle weakness
- Poor appetite
- Swallowing problems
- Salivation or vomiting
- Lameness, stiffness
- Walking with a curved back
- Fever

Work-up:

- Blood (including CBC, CMP, CPK, thyroid levels)
- Urine
- Muscle biopsy

Treatments:

- Immunosuppression with oral steroids (not all affected dogs respond to it)
- Muscle strengthening modalities including physical therapy, swimming
- Supplements, herbs

Research donations:

The following account was opened to support Prof. Mandigers work through the DCK (German Kooikerhondje Club with an initial 10.000,00 Euro donation):

BIC INGB NL2A

IBAN NL29 INGB 0007.5196.17

Recipient: Vereniging Het Nederlandse Kooikerhondje

Any amount is appreciated.

Who you should contact:

To report directly to the VHNK registry, email Esther de Leeuw (e.deleeuw@kooikerhondje.nl) or Carla Snels (carla.snels@wxs.nl)

For questions regarding symptoms or any of the information in this article, please contact Susanne Martin at susannepkmartin@gmail.com

Yours, Susanne Martin (Health and Genetics)