This article was written by Dr. Prof. Peter Friedrich, president of the VDH and ADRK show and IPO judge, and is being published with permission of the VDH and the ADRK.

Dear Rottweiler Friends,

The article on JLPP sets out in detail what is in this severe disease, how it is inherited, and how and why it must be fought. Although it is hoped that the disease is not yet widespread, complete testing of all breeding animals must continue. It is expressly emphasized that most adult dogs are carriers of the disease, even if they can not fall ill, because the disease had shown at a young age.

As of November 1, 2016, breedings may only be carried out with animals that have been tested for JLPP. This means that of all the animals that are to be found in breeding use, an oral mucosa sample must be taken and evaluated, and the result must be entered in the pedigree. Assay vials can be requested at the office. A sample (in duplicate) must be taken by an LG-breed warden, breeding judge or veterinarian. Together with the completed examination orders, to be found on the ADRK websit (forms), send the sample to the laboratory Laboklin for investigation.







After presentation of the result, a copy will be sent along with the pedigree of the dog upon the entry to the office. Before mating, the owner of the dogs involved must access the pedigree to verify that each pairing is permitted in the future.

—ADRK Executive Board

# JUVENILE LARYN-GEAL PARALYSIS AND POLYNEURO-PATHY (JLPP) IN ROTTWEILERS



## by Peter Friedrich

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#### 1. WHAT IS JLPP?

The symptoms of Juvenile Laryngeal Paralysis and Polyneuropathy (JLPP) have become apparent in Rottweilers in recent years. I would like to begin with an outline of the symptoms that are characteristic for this disorder and its typical progression. I will start by explaining where the term itself comes from. Neuropathy is a word used to describe a severe disorder of the nervous system. The prefix "poly" indicates that neuropathies can affect many different parts of the body at the same time. In a healthy body, nerve structures control muscles and perform a range of complex additional functions. In its fully expressed form, polyneuropathy causes control over the muscles to be lost to an ever-greater degree until it is ultimately lost entirely. Pronounced symptoms of paralysis of the larynx and the surrounding tissue comprise the second component of JLPP. The onset of the disease is typically marked by breathing problems in affected dogs, especially during physical exertion or when they are excited. Dogs may cough when eating or drinking, and the sound of their bark often changes. This is followed, somewhat later, by a loss of coordination in the hind limbs and subsequently also the forelimbs. These symptoms worsen until the dog is unable to move. The disorder typically appears either shortly after weaning age or in somewhat older juvenile dogs. The risk of misdiagnosis cannot always be eliminated.

The disease causes immense distress to affected dogs. It is inevitably terminal; no cure exists. We can consider ourselves lucky, that its appearance can be avoided with absolute certainty. This situation represents the best possible scenario for everybody seeking a Rottweiler puppy.

#### 2. WHAT IS THE GENETIC BASIS FOR THE CONDITION?

Juvenile Laryngeal Paralysis and Polyneuropathy (JLPP) in Rottweilers is a hereditary disease. Its mode of inheritance is monogenic autosomal recessive. It is caused, in other words, by a mutation in a single genetic locus which is not located on the chromosomes deciding a dog's sex (so not on the X or Y chromosomes). The concept of recessiveness relates to the natural laws which determine whether genes trigger disease symptoms or not. In order to understand how this works, we need to remember that every inherited trait we see in a dog that is controlled by a single gene goes back to two alleles. These are alternative forms of the same gene, one passed down from the dog's father and one from its mother. In the illustrations below, I will show how the genes behind monogenic autosomal recessive disorders are inherited and expressed. Many other modes of genetic inheritance following entirely different principles also exist, but need not be explored here; as the focus of this article is on JLPP, aspects of genetics not relevant to its transmission can be ignored here. The following genetic constellations can occur in any dog, regardless of its sex.



Type A individual = homozygous unaffected genotype: the dog's genetic make-up is homozygous unaffected ("completely clear"). Neither allele has the unfavourable mutation. In layman's terms: the dog has two good alleles. This dog will never develop JLPP. The more dogs in the population which are like this, the better.



Type B individual = carrier: the dog's genetic make-up is heterozygous. One allele is free from the unwanted mutation, and one allele has it. In other words: the dog has one good and one bad allele. This dog is a carrier of the disorder, but it will never develop JLPP. This is where the recessive heredity of the disease proves fortunate: only dogs with two mutant alleles can develop the disease. Dogs with only one "bad allele" are spared this outcome. In the context of a well-considered breeding program, gradually reducing the incidence of this gene constellation over generations is desirable. However, its presence is not of concern as long as an adequate number of homozygous unaffected dogs are available in the breeding pool.



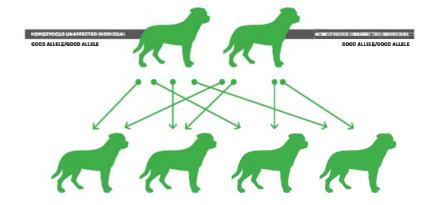
Type C individual = homozygous affected genotype: the dog's genetic make-up is homozygous affected. Both alleles carry the unfavourable mutation. This dog, in other words, has two bad alleles. This dog will develop JLPP. It is absolutely imperative that breeding programs ensure this gene constellation cannot arise.

The importance and relevance of genetic information for breeders becomes evident when we look at what happens when individuals with different genetic characteristics are mated. We can begin our exploration of this by remembering that Type C individuals with homozygous affected alleles never enter the breeding pool because they die at a young age.

#### Pairing example 1

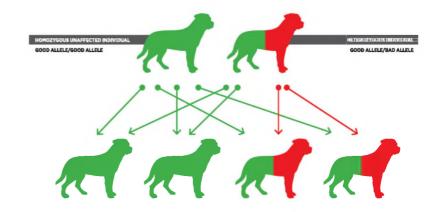
homozygous normal X homozygous normal

When two homozygous unaffected ("completely clear") animals are bred, the situation is crystal clear: none of their offspring can develop JLPP. Each son and daughter will also be homozygous normal.



### Pairing example 2 homozygous unaffectedt Xt heterozygous

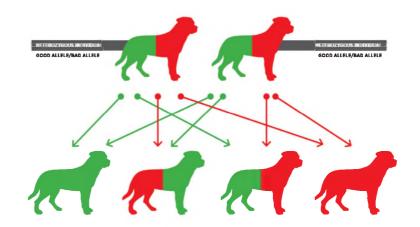
Pairings between homozygous unaffected ("completely clear") animals and heterozygous "carrier" animals also lead to offspring which are not susceptible to JLPP. If such pairings are frequently repeated, half of the offspring will typically be homozygous unaffected ("completely clear") and the other half will be heterozygous (carriers).



#### Pairing example 3

heterozygous X heterozygous

Mating two heterozygous animals (carriers) must be avoided at all costs. It can lead to offspring which develop JLPP. In large samples, it can be expected that 25 percent of the offspring resulting from such pairings will develop the disease, 50 percent will be carriers and 25 percent will be homozygous unaffected ("completely clear") dogs.



## 3. WHAT WILL HAPPEN IF THIS IS IGNORED, AND WHAT ACTION MUST ROTTWEILER BREEDERS TAKE NOW?

Fortunately, a reliable DNA test has come onto the market. Discovering whether a dog's genetic make-up is homozygous unaffected ("completely clear"), heterozygous (a carrier) or homozygous affected is now relatively straightforward and inexpensive. This test has demonstrated that the gene responsible for the disorder is present in our Rottweiler population: it has been shown that carriers exist. We don't know how many fatal cases of the disease have already occurred. Attempting to trace the problematic genetic heritage of dogs back to particular ancestors does not represent a fruitful approach to the issue: the best way forward lies in the adoption of genetic testing. At this point, let us imagine, for the sake of argument, a scenario in which very few Rottweilers were carriers. If that were the case, some people might reject the option of testing on cost grounds as well as for other reasons: measures to counteract a rare disease might strike them as disproportionate. In their innermost selves, they might also harbour deep-seated fears that they would no longer be able to breed from one of their most successful dogs without restrictions. A few people might handle the resultant stress by going into denial and refusing to engage with the issue or find out more about it. Knowing nothing about the disorder, they would then not need to factor it into their thinking. If you are in any way tempted to react in this way, please consider changing your attitude, even if you find this difficult, for the sake of the Rottweiler. Please wake up to the fact that responsible, ethically justifiable and legally compliant dog breeding must necessarily rest on more solid foundations. The risk of the mutant gene slowly but steadily spreading throughout the population is unacceptably high. Over years, the number of carriers would rise higher and higher, and sooner or later the number of fatal cases of the disease would also rise drastically. We know all this. And we must draw the logical conclusions from what we have learned.

From now on, or, more accurately, after an appropriate transition period, to allow time for the necessary tests to be carried out, every single breeder may only breed from dogs that have been identified as homozygous unaffected ("completely clear") or as carriers. Carriers may only be bred with homozygous unaffected ("completely clear") animals. Homozygous unaffected ("completely clear") dogs may be bred with both homozygous unaffected ("completely clear") dogs and with carriers. The procedure specified here ensures that NO more Rottweilers will develop JLPP.



To be clear: I want to stress here that it would not be at all wise to breed only from homozygous normal ("clear") dogs and bitches. If that were to happen, genetic diversity would be severely reduced and it would be virtually impossible to stave off the negative consequences of inbreeding.

The ADRK Board has, quite correctly and after consultation with the breeding committee, now adopted a resolution to this effect. It will go on to produce a list of authorized genetic laboratories of assured quality. Independently of any club regulations, the same duty to take action can also be derived from the German Protection of Animals Act which each and every one of us is bound by. But let this be clear: our desire to breed healthy, sociable, energetic, and fun-loving Rottweilers that are true to type comes from the bottom of our hearts. It is not something we do only for the sake of complying with legislation.

I perceive the same ethical duty to adopt precisely this approach in every country where Rottweilers are bred, be that Germany or any other nation. Diseases do not respect national borders. The suffering of animals does not stop at national borders. Friends of the Rottweiler across the world – on every continent and in every country – are challenged to act on this issue. Nothing and nobody can free any breeder from this duty. Whether the relevant local breeding regulations already contain provisions dealing with JLPP matters not a jot in this regard.

In conclusion, it may be noted that other dog breeds have already profited from the successful implementation of similar breeding programs. Several diseases with the same mode of inheritance have already been eliminated in this way, for example polyneuropathy in Greyhounds (a relatively similar form to the disease affecting Rottweilers) and progressive retinal atrophy (PRA) in Sloughis. Counter-measures against JLPP in Rottweilers are exceptionally likely to prove successful, not least because no other genetic tests are currently in routine use in their breeding.

