

Legg-Calvé-Perthes Disease

Camden's Story

By Robert McCaskill, DVM; Matt and Terri Grant

We are providing the following article to you, our Westie Wellness readers, to inform you of Legg-Calvé Perthes Disease (LCPD), a painful (and expensive) disease.

Providing blood samples to the Westie Foundation of America Biobank will enable researchers to link DNA with health issues such as Legg-Calvé-Perthes Disease. Please recall that samples need not be from pedigreed Westies. Samples can also come from a rescue dog or a dog of unknown lineage. Blood samples from which DNA will be extracted are needed from healthy dogs for the research control purposes as well as dogs with health issues.

To request a sample kit and for more information: www.westiefoundation.org/biobank.



He was the last puppy born on Christmas Eve out of a litter of three. His brother and sister before him were stillborn. We called him Dickens. His mom had previously free-whelped three litters. He had a barely audible heartbeat. It was weak but there. I went to work to ensure that his airway was clear. I started chest heart massage and mouth to mouth breathing. Finally, after many minutes he took a breath. He weighed 200 grams, a normal size westie puppy. I spent the next 3 days ensuring he was nursing and providing supplemental nourishment every 2 to 4 hours. He gained weight and with that, strength. By the time he was 300 grams, I was confident that he was strong enough to nurse and survive. His mom, Emma, has never been a great housekeeper. I assisted with warm water cotton ball wipes to ensure he had bowel movements. By day 5, Emma started doing her share. By two weeks, I knew he was going to be a smart and special puppy.

COVID had struck. Schools were closed. Our grandson, Ethan, was with his grandmother enrolled in virtual learning. On his breaks, Ethan would occupy his free time playing with Dickens.

Their favorite play was chasing a fuzzy duster that was on a pole. Dickens would chase and go for the duster over and over. Dickens also liked toys. He would bring them out of his toy box. He would play fetch until bored. Then he would lay at my feet napping. At 4 months, I noticed that one testicle was down. I watched and by 5 months, the other testicle was not going to completely descend. We knew that this fantastically smart puppy could not be shown or used in our breeding program.

I had recently been contacted by Matt Grant. Matt and his wife, Terri, had obtained a westie boy, Bentley, from us a couple of years earlier. Terri and Bentley enrolled in therapy dog training taught by Jane Fink. Bentley received AKC Certification, but then COVID hit closing hospital visits. They wanted another boy to also train as a therapy dog. Thus, Dickens went off to Matt and Terri's to join this super family. Matt and Terri now called their boy Camden. In December, Matt called me. He had just had Camden neutered. After the procedure, Camden had started dribbling urine. Camden had also started a slight occasional limp on his left rear leg. Camden's clinician stopped some of the post-surgical meds and took an x-ray. The urine dribbling slowed but did not stop. The x-ray showed no pathology, but Camden's clinician suggested a consultation with a veterinary orthopedic surgeon. One was scheduled. A visit was made with no definitive diagnosis. The surgeon suggested that Camden return for a follow-up visit in six months. This specialist could provide no thoughts about the urine dribbling.

Matt and Terri then scheduled a consultation with a veterinary internist who had a special interest in urology. A complete work up to include a CAT scan was performed. Nothing eventful was found by the specialist. The specialist prescribed some meds and antibiotics. Camden's urine dribbling improved but did not go away. The CAT scan showed some orthopedic changes in Camden's right rear hip. They referred him back to the veterinary orthopedic surgeon who scheduled a follow up consultation. X-rays now showed changes in the right femoral head. A diagnosis of Aplastic Necrosis was made. Aplastic Necrosis is the scientific term for Legg-Calves Perthes Disease (LCPD). The surgeon presented Matt and Terri with several

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surgical options with a recommendation of a total hip replacement. The surgeon recommended the hip replacement as the best option for returning Camden to full mobility. Matt and Terry decided to go with this procedure.

Camden had the surgery. He stayed in recovery for three days. He walked out of the hospital on his own. Camden's exercise was restricted for 6 weeks. Camden had a follow-up visit with his surgeon 9 weeks later. He was healing nicely. He was placed on limited exercise. By 12 weeks, he was allowed to return to normal activity. Interestingly, as soon as the surgical procedure was done, Camden quit dribbling urine. The urine dribbling has not returned. I have spoken with several specialists who have not observed this condition before. We believe that it was associated with pain because when Camden went on walks before the surgery the dribbling would start. By then, it was not as dramatic as it was initially, but it was still present.

Today Camden has returned to chasing the squirrels in the back yard. He and Bentley now enjoy long walks through the neighborhood. Terri plans to resume training with Jane Fink. The surgery went well as did the recovery.

Legg–Calvé–Perthes Disease (LCPD)

By Dr. Robert McCaskill, DVM

My experience with LCPD started with a West Highland, Kiltie. She came into our life while we were stationed in Germany. She was a fantastic puppy. We were then posted to California with the golden retriever and Kiltie. At 11 months of age, Kiltie started limping on her left rear leg. X-rays showed femoral head flattening with changes in the acetabulum of the hip. The diagnosis was LCPD. I reached out to Dr. Bob Rooks, a noted veterinary orthopedic surgeon in Fountain Valley, CA. We scheduled Kiltie for a consultation with potential for a femoral head ostectomy which means removal of the femoral head. Bob allowed me to assist with the surgery.

The surgery and recovery were uneventful. After a couple months, Kiltie was running with me and the golden. For 30 years, LCPD had not shown up again in our breeding program until it showed up with Camden. Discussions with both veterinary and human orthopedic surgeons indicated that there was no evidence of a genetic component and that perhaps environment played a part.

But now Cornell University's College of Veterinary Medicine has identified a possible gene on Chromosome 6. If this can be substantiated, it would benefit not only our breed but also potentially humans. Dr. Rory James Todhunter is the research team lead and veterinary orthopedic surgeon. His article on LCPD follows. The research team believes that LCPD may be an autosomal recessive genetic disease. If this is true, they believe that they can develop a genetic test. That means our Westie World would have another tool to assist us in potentially eliminating this disease and pain from our breed.

(Thanks to Anne Sanders & Steve Schroffel for edits, formatting and compiling this article.)

Legg–Calvé–Perthes Disease

*By Rory J. Todhunter BVSc, MS, PhD, DACVS
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THE GOAL

Legg–Calvé–Perthes disease (LCPD) is a juvenile-onset, secondary collapse of the femoral head due to death of the cells that produce its cartilage and bone – a process known as osteonecrosis (bone death). The result is hip deformity and premature hip osteoarthritis or degenerative joint disease and is most seen in small breed dogs of both sexes. The trait is also called avascular necrosis (loss of blood supply) of the femoral head.

Our long-term goal is to develop a genetic test which can be used to make informed breeding decisions in susceptible West Highland White Terriers. Breeding dogs not carrying the mutations will reduce, and hopefully eliminate, the disease from the breed. The current state of the art to find a causative genetic region or locus for this disease is to undertake a molecular genetic screen across the entire genome of cases and controls. The goal is important because (as illustrated by the preceding stories), LCPD causes pain and lameness, it usually requires surgery (removal of the femoral head and neck or total hip replacement) to successfully eliminate the hip pain. This is a risk to the affected dog because both procedures are invasive, painful, and require a long convalescence, in addition to the substantial cost to the owner.

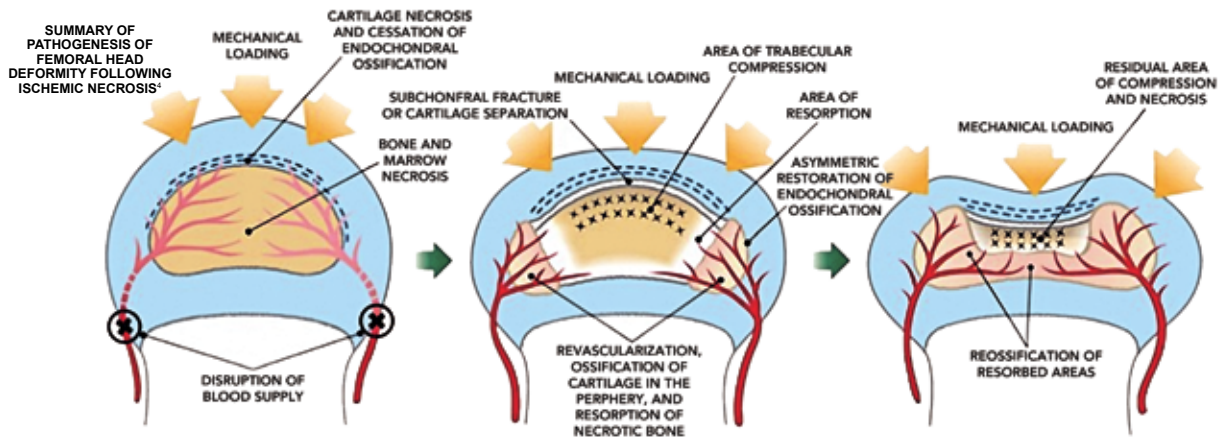
THE TRAIT

Legg–Calvé–Perthes disease was first described in adolescent humans by the English, French and German physicians, Legg, Calvé, and Perthes, respectively, in 1912. In humans, boys have a higher incidence. In dogs, there is no sex bias. Clinical

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LEGG-CALVÉ-PERTHES DISEASE

Summary of Pathogenesis of Femoral Head Deformity Following Ischemic Necrosis⁴



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Figure 1

signs of predominantly unilateral hip pain and lameness appear in affected dogs between 4 and 11 months of age.

Disruption to the blood supply of the femoral head in the hip joint socket (*acetabulum*) produces hypoxic (*low oxygen*) injury to the developing femoral head.¹ (See Figure 1). The blood supply to the femoral epiphysis enters the femoral neck at the joint capsule reflection and runs up the neck into the epiphysis (*top of the femur*). Anatomic vascular studies in susceptible small breed dogs demonstrated that the femoral epiphyseal blood supply is less protected, and therefore vulnerable to trauma or occlusive (*blocking*) pressure, in the region around the growing cartilage in the femoral neck when compared to the vascular architecture in non-susceptible mixed-breed dogs.² Other remarkable similarities between LCPD in children and small breed dogs, include low insulin-like growth factor-1 (*IGF-1*) levels, generalized small artery caliber with reduced arterial function, and a hyperactive personality.³ Insulin-like growth factor-1 is the hormone that acts locally to stimulate the cartilage cells to grow larger and thus make the whole organ larger.

GENETICS

In our preliminary unpublished study, we searched for associated regions of the genome that might affect risk for LCPD using samples from several small breeds of dog. We

have discovered a genetic region that seems to associate with LCPD but not at a statistically significant level for a genome wide analysis. We propose to extend our preliminary studies, to discover the first genetic marker locus associated with this important disease in small breed dogs. Further, we will investigate the genetic variants identified within the associated region to determine the candidate causal mutation(s). The DNA samples in the Westie Foundation of America Biobank and other collected DNA will be a critical next step to continue our research. Your samples will greatly help us to genotype many more cases and controls of West Highland White Terriers and closely related breeds.

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