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POODLE Update

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EYE DISEASE RESEARCH

Advancements Depend on Input
from Breeders & Owners

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RESEARCH OF POODLE EYE DISEASES NEEDS SUPPORT FROM BREEDERS & OWNERS

Eye diseases in Poodles have long been a health concern. When Poodles joined the [Canine Health Information Center \(CHIC\) program](#) in 2004, eye examinations were one of the initial requirements across all three varieties for CHIC certification. Despite efforts to encourage eye tests and advise breeders to selectively breed against eye diseases, one expert notes an ongoing steady increase in Poodle eye diseases.

Gustavo Aguirre, VMD, PhD, professor of medical genetics and

ophthalmology at the University of Pennsylvania School of Veterinary Medicine, says, “Vision problems still affect far too many Poodles. Compounding efforts to identify gene mutations, we have a shortage of DNA samples from all varieties of Poodles. It takes a commitment from breeders and owners working with researchers to help advance discovery of causative gene mutations. Then, it’s up to breeders to selectively breed dogs that will not produce offspring with eye diseases.”



PROGRESS REPORT ON MINIATURE & TOY POODLE EYE DISEASES

The Poodle Club of America Foundation is helping to fund a six-year genetics study that began in 2014 at the University of Pennsylvania. The research, led by Dr. Gustavo Aguirre, is focused on helping to advance understanding of optic nerve hypoplasia/micropapilla and cataracts in Miniature and Toy Poodles.

Optic nerve hypoplasia (ONH) is a non-progressive congenital defect causing blindness or severely impaired vision. Miniature Poodles have a high prevalence, and a few cases have been reported in Toy Poodles. Considered a rare genetic defect, ONH affects one or both eyes and occurs when the optic nerve fails to develop normally. Disease severity is based on the extent of damage to the optic nerve.

Originating from the optic disc at the back of the eye, the optic nerve is made up of nerve fibers that connect retinal cells with visual centers in the brain. The optic disc appears grayer and abnormally small in dogs with ONH due to fewer nerve fibers. This means that the brain receives fewer signals for light reception.

In the ONH study, the researchers looked at the pedigrees of 43 Miniature Poodles, nine of which were affected by ONH in both eyes. They found a possible common founder, though none of the candidate genes tested correlated with ONH. The study [was published in the January 2020 issue of *Veterinary Ophthalmology*](#).

Meanwhile, Miniature Poodles also are susceptible to micropapilla (Mp), a condition similar to ONH in which dogs have abnormally small optic discs. Unlike ONH, Mp usually is not associated with a visual defect, implying that Mp could be

a less severe form of ONH or a totally different disorder.

“I feel that ONH and Mp are part of a continuum,” Dr. Aguirre says, noting that the two conditions often appear in the same Poodle pedigrees. “I speculate that ONH is a disease caused by genes at two separate loci, but that research needs to continue in order to prove or disprove the hypothesis.”

Dr. Aguirre advises breeders not to breed dogs with ONH; however, he says there is not enough information to preclude Poodles with Mp from being bred. For both eye diseases, puppies can be examined as early as 6 to 8 weeks of age to identify the conditions before going to new homes. Breeders are encouraged to have eye examinations of their litters.

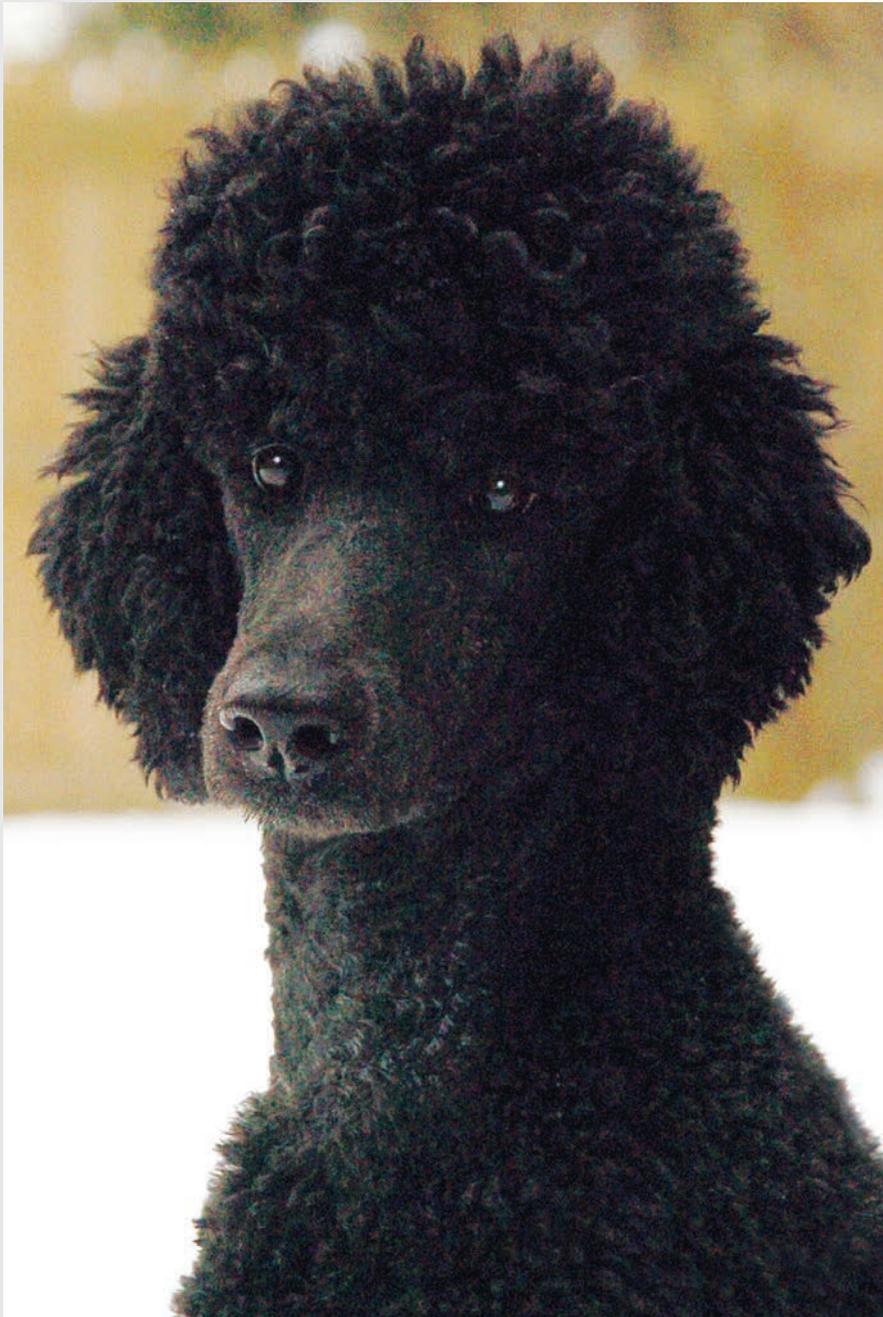
Hereditary cataracts can be challenging because they typically do not appear until a dog is around 4 to 5 years of age, oftentimes after already receiving eye clearances from health testing. In Toy and Miniature Poodles, dogs are born with normal lenses that gradually lose their transparency. Cataracts can involve the nucleus, or central part, of the lens, as well as the lens cortex, or more peripheral layers. The rate and progression of cataracts vary.

“When older Poodles present with cataracts, there is no way to know if it is due to aging or heredity unless those dogs have records of previous exams dating to their younger years,” Dr. Aguirre says. “We would like to search for the mutation in Poodles, but we need more samples. Cataracts can be caused by other things, such as progressive retinal atrophy, so we need more samples to account for these possibilities.”

A renowned eye disease researcher, Dr. Aguirre discovered the mutation for the progressive rod-cone degeneration form of progressive retinal atrophy (*prcd*-PRA) in Toy and Miniature Poodles in 1998. DNA testing for *prcd*-PRA is a required test for CHIC certification for these varieties.

In efforts to better understand the genetics and treatment options for eye diseases, [the Poodle Club of America Foundation is helping to fund eye disease research](#). Currently, the Foundation is supporting genetic research of polymicrogyria and day blindness/retinal degeneration in

Standard Poodles, as well as studies of optic nerve hypoplasia/micropapilla and cataracts in Miniature and Toy Poodles. The goal is to reduce the incidence of eye diseases in all varieties of Poodle through education and development of genetic tests.



Born with the neurological disease polymicrogyria, “Celie,” a Standard Poodle, was visually impaired and had partial complex seizures contributing to emotional and behavioral problems.

NEUROLOGICAL DISEASE PMG & VISION PROBLEMS

There was nothing unusual about the newborn black Standard Poodle at first. Like her littermates, she felt and sniffed her way around the whelping box. When their eyes began to

open, she continued to find her way around by smelling and feeling.

Curtisy Briggs of Malvern, Pennsylvania, adopted the puppy in December 2002 at 10 weeks of age knowing she was blind. “I named the puppy ‘Celie’ after the protagonist in ‘The Color Purple.’ I hoped that the puppy would be strong and overcome adversity like Celie did,” she says.

The breeder had taken the puppy to a veterinary ophthalmologist, who determined her eyes were normal. The problem appeared to be neurological and have to do with visual processing in the brain, though the cause of blindness was a mystery.

“I noticed quickly that Celie was not completely blind,” Briggs says. “She seemed to see shapes and followed my other dogs unerringly in a chase. I sometimes saw her avoid obstacles in her path, and she would sit and look into the bright sunlight as though she was watching the trees or sky. I wondered if her eyes could see, but her brain could not organize the picture right.”

Unbeknownst to Briggs at the time, Celie suffered from the neurological condition polymicrogyria (PMG), which often but not always affects vision. The puppy’s eyes had normal vision. The optic nerves that carry visual signs to relay centers in the brain, which control pupil reflexes, eye movement and some visual processing, were normal, as were the relay centers. The abnormality was at the visual cortex. All that Briggs knew was that Celie was

POODLE OWNERS & BREEDERS CAN HELP ADVANCE EYE DISEASE RESEARCH

Breeders and owners of all varieties of Poodle are encouraged to submit DNA from dogs affected with eye diseases, including polymicrogyria in Standard Poodles, to Dr. Gustavo Aguirre and his team at the University of Pennsylvania. To submit samples, please contact Dr. Aguirre's lab at 215-898-9426 or by email at lmelnyk@vet.upenn.edu.

Researchers at the University of Missouri are seeking DNA samples of Standard Poodles diagnosed with polymicrogyria. "The more samples we have, the more powerful our studies will be and the more likely we will be to find an answer," says Dr. Dennis O'Brien. "It is critical, however, that the diagnosis is confirmed by a veterinary neurologist, as there are other diseases that can cause vision problems." Please contact Dr. O'Brien at ObrienD@missouri.edu to learn how to participate.

basically blind and had somewhat odd behavior.

Located near the rear of the outside surface of the brain, the visual cortex processes visual signals into recognizable objects. The brain's surface, or cortex, is normally covered in large folds called gyri. However, in dogs with PMG, the cortex has many excessive small folds, or gyri, that disrupt normal connections within the cortex.

Functioning of the temporal cortex, located in front of the visual cortex, may be severely affected as well. The temporal cortex is involved in emotional responses, which is why some dogs with PMG have personality issues and seizures.

Celie was exceptionally affectionate and craved closeness with Briggs when they were together in the house, but outside in the yard was another story. Celie often ran in circles, which Briggs thought was an orienting mecha-

nism, but sometimes the circling became frantic and she refused to listen or be caught. "It was almost as though she was in the grip of some urgent force compelling her to ignore everything around her," Briggs says.

The obsessive circling progressed, and Celie was not able to stop herself as she ran to near collapse. Briggs stopped playing in the yard with Celie because any excitement seemed to bring on the rampant circling. She referred to this as Celie's "fugue state," a condition in which she seemed unaware of her surroundings and often slammed into things she normally avoided.

Briggs observed that Celie sometimes smacked her lips and slightly shook her head in situations that made her nervous. Calming medication helped for about one year but eventually became ineffective. One day when Celie was almost 5 years old, she lunged at Briggs biting her hard

on the leg. The Standard Poodle's behavior continued to worsen.

"Once I turned around to find Celie a few yards away hunched over and about to spring toward me," Briggs says. "For the first time in my life, for a split second, I was afraid of my own dog."

Briggs decided to take Celie to a Poodle eye clinic where Dr. Aguirre, whom she had consulted about other Poodles, was going to perform eye examinations. Dr. Aguirre immediately diagnosed Celie's problem as coming from the brain, rather than being a vision problem, and told Briggs he had seen a few Standard Poodle puppies with a type of neurological blindness known as polymicrogyria. He had written an article describing four affected dogs that was [published in 1994 in *Progressive Veterinary Neurology*](#).

"The first affected dog I saw was a white male Standard Poodle," Dr. Aguirre says. "The dog had bizarre visual behavior. He could not see, but he also was not totally blind."

Dr. Aguirre has since examined quite a few Standard Poodles with PMG and described a range of phenotypes. "Some dogs are able to see more than others," he says. "Most, but not all, have a form of hydrocephalus, or fluid buildup in the cavities deep within the brain. We see one to two cases a year. There are undoubtedly more cases, though most affected dogs are likely euthanized as puppies. It also is likely that dogs that are mildly affected are just not diagnosed unless a magnetic resonance imaging (MRI) study is done."

Believing that Celie had PMG, Dr. Aguirre referred Briggs to a veterinary neurologist at the University of Pennsylvania, who confirmed the diagnosis based on MRI. The specialist also diagnosed Celie as having partial complex motor seizures, which accounted for the unusual behaviors of circling, lip smacking and head shaking.

Ironically, the calming medication that Celie took lowered her threshold for seizures. She had to be weaned off the medication, as every seizure caused more brain damage. Without the medication, Celie was increasingly wild, and she had begun spinning around the house too.

“I knew Celie had to be suffering in ways I could not even fathom and that my other dogs were paying a price for living with her erratic behavior,” Briggs says.

When Celie was almost 5 years old, she was the oldest dog known to have PMG. Unfortunately, the disease ultimately won, and Briggs made the difficult decision to have her euthanized. At least there was solace for Briggs knowing that Celie had brought attention to the disease and that her DNA had gone to genetic research.

“It is so important to get a genetic test because breeders are producing affected puppies that they attribute to bad luck,” Briggs says. “They euthanize affected puppies and then repeat the breeding, producing more affected dogs. Without a test, there is no way to know which puppies are affected with the specific disease. Since abnormalities vary so much, PMG can be overlooked as a cause. Not all dogs with PMG are blind, not all

have motor problems, and not all have behavior or emotional problems or seizures. Many of these dogs with PMG are likely going undiagnosed.”

Veterinary neurologist Dennis O’Brien, DVM, PhD, the Chancellor’s Chair in Comparative Neurology at the University of Missouri College of Veterinary Medicine,



has been studying PMG since 2007. He knows firsthand the challenges of living with a dog with the disorder. “Oliver,” a white male Standard Poodle, came to live with him in 2008 after being diagnosed with PMG.

Unfortunately, Oliver, who was more severely affected than Celie, lived with Dr. O’Brien a short time. “He was a sweet dog, but it was tough for him to manage with poor vision,” he says. “He was getting injured too frequently, so I made the decision to euthanize him.”

Since Celie’s case, more Standard Poodles, some even older than Celie with less severe PMG,

have been diagnosed. “As I did with Celie, owners may struggle for years without knowing what the problem is,” Briggs says.

Describing the genetic work at the University of Missouri, Dr. O’Brien says, “The initial gene mapping study in Dr. Gary Johnson’s lab found a locus more common in affected dogs than in the population at large. This locus has been difficult to work with, but we have investigated that area extensively as well as performed searches of whole genome sequences we’ve acquired. We have not found the culprit yet. We hope to someday make PMG a disease of the past.”

DAY BLINDNESS/RETINAL DEGENERATION IN STANDARDS

In 2013, Wendy Grogan of Hackettstown, New Jersey, had a litter of five Standard Poodle puppies — one white female and four black males. She decided to keep the white female. It soon became apparent that the Poodle she named “Sugar” walked into everything in her path when she was outside, yet she was fine in the house.

“My son said, ‘She’s either very stupid or blind,’” Grogan says. “I knew she wasn’t stupid. I watched her carefully for the next week, and then I emailed Dr. Aguirre whom I knew from helping my mother bring dogs to his eye clinics. I told him that it almost seemed like she was blind but only when she went outside.”

Dr. Aguirre told Grogan he had examined another Standard Poodle, coincidentally named

“Sugar,” in the 1980s who was one of three puppies in a litter of five having difficulty seeing in bright light. “It is rare for dogs to have difficulty seeing in just bright light,” Dr. Aguirre explains. “It is more common when there are retinal problems for dogs to have trouble seeing in dim light, indicating problems with the rod retinal receptors. When bright light is a problem, it means there are problems with the cone retinal receptors.”

Dr. Aguirre did not hear of another case until early 2000 when he began examining other Standard Poodles with day blindness. When he learned about Grogan’s Sugar, it was an opportunity to study the family. Examining the littermates, sire and dam, he found only Sugar was affected.

When the first Sugar was diagnosed, an electroretinogram (ERG) was used to record the retina’s electrical response to light or a veterinary ophthalmologist would check the retina of the eye after it had been removed. Today, optical coherence tomography (OCT) provides a high-resolution method of examining anatomically a dog’s retina, though a dog must be anesthetized for the procedure.

Meanwhile, analysis of the DNA of Sugar and her relatives allowed Dr. Aguirre and Karina Guziewicz, PhD, research assistant professor of ophthalmology, to identify the gene mutation. Subsequently, a DNA test was developed for Poodle day blindness/retinal degeneration (DB/RD). The test is available from the diagnostic testing lab Wisdom Health, formerly Optigen.

Thus far, DB/RD has only been reported in Poodles. It is a progressive condition that affects the cone and rod retinal receptors, rendering some though not all Poodles totally blind initially. “It is a variable disorder,” Dr. Aguirre explains. “Some affected dogs at 2 years of age are indistinguishable from dogs with end-stage progressive retinal atrophy, yet in other dogs the retina remains clinically intact for several years.”

Sugar was adopted at 4 months of age by a veterinary technician, Cristie Nielsen of Oceanport, New Jersey, who knew about the vision problem and hoped a treatment would become available. Unfortunately, Sugar’s vision worsened.

“When Sugar was about 1 year old, I noticed that she had started bumping into things indoors,” Nielsen says. Despite being virtually blind, Sugar, now 6 years old, romps with Nielsen’s male Miniature Poodle, goes for walks and leads a full life.

Vision problems like day blindness/retinal degeneration and PMG vary considerably among individual Poodles. The good news is that with help from researchers and diligent breeders who regularly health test their breeding stock, some of the hereditary eye diseases may one day no longer be a problem. ■

Purina thanks Pat Forsyth, vice president of the Poodle Club of America Foundation board of directors, for helping us to identify this topic for the *Poodle Update*.

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