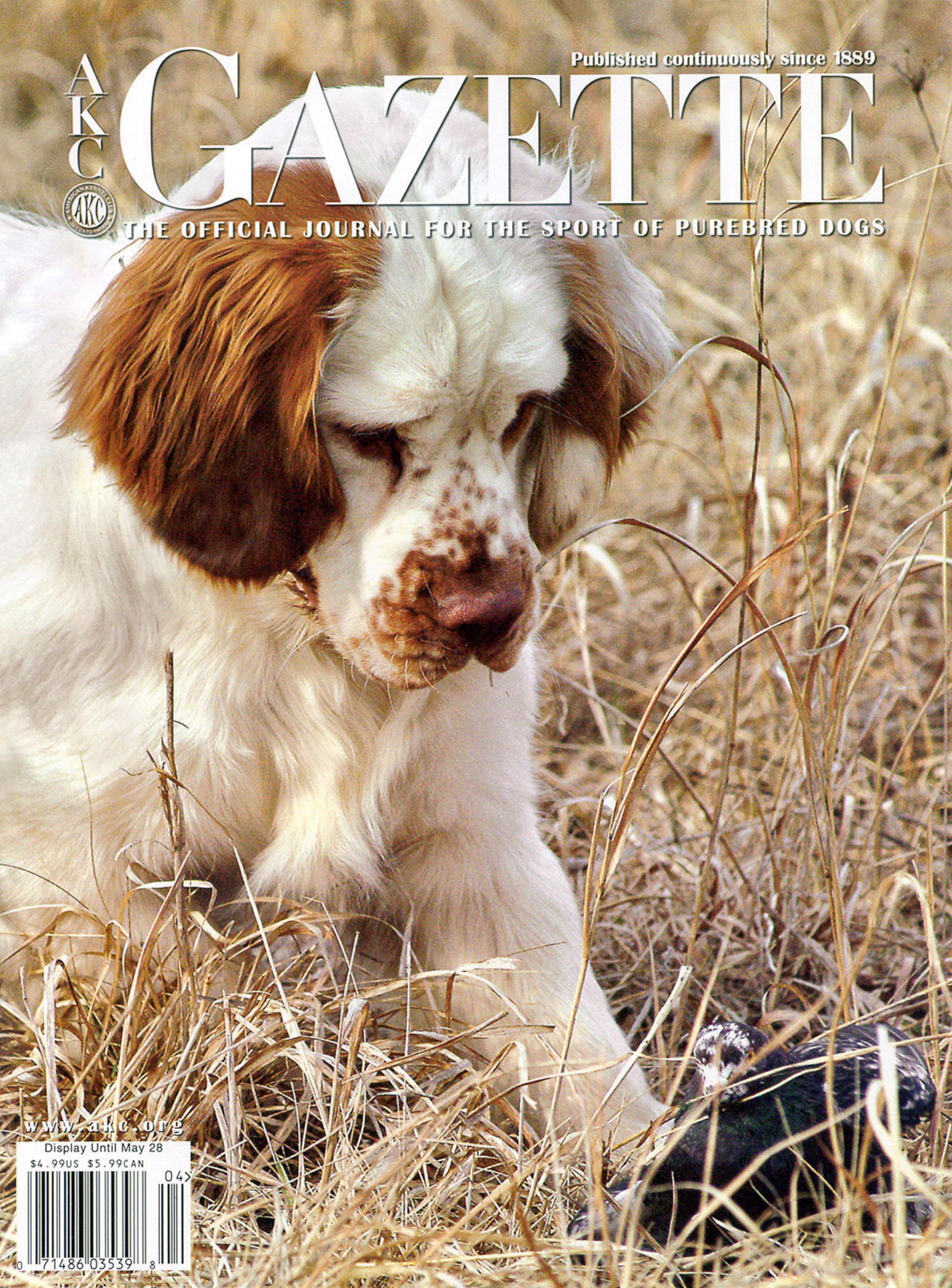




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# Heartbreak and Hope

**A**t the Animal Eye Clinic, in Wilton, Connecticut, the exam room is dark. Charles M. Stuhr, DVM, DAVCO, uses a hand-held microscope to peer into the eyes of Misty, a toy Poodle. He has gently coaxed information from her worried owner, Eric Breuning, explaining, "She can't tell us whether she sees or not, so your description of her behavior is an important tool in her assessment." Softly clicking his tongue to get Misty's attention, he waves his fingers in front of her eyes, eliciting little, if any, response.

The exam completed, Misty is free to wander the room. Breuning thinks that she still has some sight, although she had recently fallen down a flight of stairs. He is reluctant to think of his dog as blind.

Stuhr dims the light again and asks Breuning to call Misty. The Poodle turns her head frantically at the sound of her master's voice, but is unable to locate him. For Dr. Stuhr, who sees about 5,000 patients a year, it's another dramatic example of how well dogs can mask their blindness, even from the most astute owner.

Like most owners whose dogs are diagnosed with cataracts, glaucoma, or progressive retinal atrophy (PRA), Bruening will undergo classic stages of denial, grief, and loss. Depending on the diagnosis, Misty will experience varying degrees of distress. Both owner and dog will have to adjust to the vicissitudes of canine blindness, and Misty's breeder will face difficult decisions about the future of the line.

## GLAUCOMA

Striking one in 200 dogs, glaucoma is among the most frequent causes of canine blindness. As Ann Warren, of St. Paul, Minnesota, knows all too well, it is also the most agonizing. In January 2003, Chase, her 4-year-old Chinese Shar-Pei, started to tremble. Disoriented, he staggered off the edge of the couch, squinting and pawing at his eyes. Ann's frightened dogsitter rushed him to an emergency vet.

Later, a veterinary ophthalmologist gave Warren devastating news: Chase had glaucoma. His intraocular pressure (IOP), normally below 20, measured 47 in his left eye; his right eye spiked at 62. Humans with high pressure describe the feeling as an excruciating headache.

Elevated IOP is the most concrete symptom of glaucoma, a frustrating and complex disease with a poor prognosis. It occurs when the normal outflow of aqueous humor—the fluid produced by the ciliary body behind the iris—is blocked. Ordinarily, this fluid, which shapes and stabilizes the eye, seeps out through the pupil and drains at the junction of the cornea and iris (the iridocorneal cleft). In glaucoma this flow becomes obstructed, first in one eye then, eventually, the other. Pressure builds and damages the optic nerve, resulting in pain and impaired vision.

Early diagnosis is critical to prolonging sight. If the dog can see at the time of exam, he will be treated with an aggressive regimen of eye drops and medicines to reduce pressure and fluid production, improve outflow, and help drainage. These include

**Knowledge of the causes and treatment of glaucoma, cataracts, and PRA will help you cope with these relentless hereditary diseases.**

**BY SUSAN LENNON**



ENGLISH COCKER SPANIEL/ISABELLE FRANCAIS FOR AKC

Timoptic, Neptazane, and Zalatan. Hospitalization, various surgeries, and intravenous drugs may also be recommended, with the goals of restoring or preserving vision in the affected eye and staving off progression in the good eye. Medicines required after the acute episode are expensive and labor-intensive to administer at home. Treatment may preserve eyesight temporarily, but glaucoma relentlessly and painfully progresses toward blindness—usually within two years.

In the latter stages, destroying the ciliary body with an antibiotic injection (ablation) is a technique that can control pressure, but it causes blindness in a dog who may still have sight, and it

is not a lasting cure.

Permanent pain-relief becomes the ultimate therapeutic goal. Caroline D. Levin, RN, author of *Living with Blind Dogs: A Resource Book and Training Guide for the Owners of Blind and Low Vision Dogs* (Lantern Publications, 2001), recommends surgically removing the eyeball and sewing the eyelid shut (enucleation) or, optimally, taking out the contents of the eye and implanting a silicone prosthesis (evisceration). “Each remedy has its advantages and disadvantages, and evisceration, while the most cosmetically pleasing, may not be indicated based on breed, age, health, or history. It’s a very difficult decision for a care-

*Breeding animals should have their eyes checked annually, even after they are retired.*

## Coping With Blindness

Ann Warren, whose Shar-Pei has glaucoma, says you need to keep your perspective. "This is a real mourning process; I lost the dog I had. Glaucoma robs you and your dog of more than just sight, yet it's not a death sentence." Caroline Levin's book *Living with Blind Dogs* has helped her cope.

Kristi Murdock, whose standard Poodle Murphy has cataracts, agrees: "Although of course I wish that the puppies didn't ever have cataracts, I have come to realize how resilient dogs can be about sight impairment, and to recognize that, although this was a devastating experience, at least the dogs are able to lead happy, relatively normal, and otherwise healthy lives."

An Internet group at [groups.yahoo.com/group/blinddogs/](http://groups.yahoo.com/group/blinddogs/) provides support and information for more than 1,900 members. ♦

giver, but the dogs are so much happier without the diseased eye—free from pain, they become like puppies again."

Chase still has some sight, and Warren is treating him with both conventional and alternative modalities.

In the United States, 22 breeds have heritable glaucoma at a rate of two percent or higher. Breeds most susceptible include the Cocker Spaniel, Basset Hound, Beagle, Boston Terrier, Chow Chow, Norwegian Elkhound, Samoyed, Shar-Pei, Shi Tzu, Siberian Husky, and Wirehaired Fox Terrier. The incidence for Cocker and Basset is over six percent.

For every breed except the Beagle, the mode of inheritance for primary glaucoma is still not determined. As a disease of middle age, it is virtually impossible to detect until it's too late for most breeding programs. Kirk N. Gelatt, VMD, who has studied canine glaucoma for more than 30 years, says that affected dogs should not be bred, and that breeders should have annual eye checks on their dogs, even after they are retired. "Find an ophthalmologist to work with, take affected dogs out of your breeding programs, keep good records, and watch pedigrees."

Genetic testing for glaucoma is a long way off.

### CATARACTS

In February 2003, Jeri Moore's Prince Tye, a 12-pound apricot toy Poodle, nearly fell down the stairs. Picking him up, Jeri was startled to see that his eyes were cloudy. Tye, age 7, was diagnosed with bilateral cataracts and recommended for lens-removal and -replacement

surgery. His prognosis was good: preservation of 60 percent of vision in his right eye and 90 percent in his left.

According to Carmen Colitz, DVM, a molecular biologist and veterinary ophthalmologist at Ohio State University, restoration of sight as the result of cataract surgery is 95 percent successful. It is also the only treatment available, and early detection and intervention are essential for positive outcomes.

Often mistakenly thought of as a film over the eyeball, cataracts occur deep within the eye. Evolved from Medieval Latin translations of ancient Arabic, the word *cataract* literally means "to break down." It's an opacity of the lens that scatters light, looks gray or white, and causes

visual impairment or blindness. Most cataracts are progressive, although the rate is difficult to predict. Some can worsen dramatically overnight, some remain static for life.

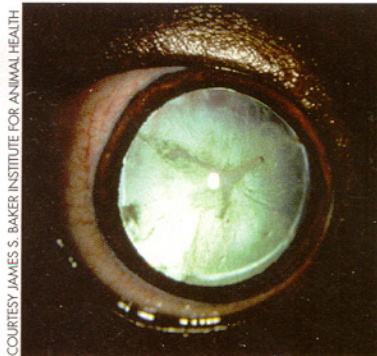
For the five percent of dogs whose surgery is unsuccessful, owners confront escalating costs, supplementary surgeries, and complications including glaucoma. Moore and Tye, who live in La Habra, California, fall into this category. Tye's care has cost more than \$5,000 and, in addition to visiting the ophthalmologist every few weeks, he faces further operations.

Often diagnosable in puppyhood, cataracts can devastate a breeding program. Kristi Murdock, of Toddville, Iowa, did everything right in 1999, when she bred her blue standard Poodle, Murphy, for the first—and only—time: "We performed all the suggested tests, and the sire is a proven dog who was also current on all testing. It was a total outcross, with an inbreeding coefficient of less than 10 percent on 10 generations."

Yet, disaster struck—5 out of the 10 puppies developed cataracts by the age of 7 months. Kristi refunded purchase prices, took one pup back on request, and remains in touch with them all.

Colitz has been studying cataracts since the mid-1990s. She says that no genes have yet been identified, but the mode of inheritance is assumed to be autosomal recessive. To be safe, she recommends that breeders neuter any affected dog, their parents, and possibly all of the affected dog's offspring. Murdock did just that with her dam and puppies.

Breeds predisposed to heritable cataracts with



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*A cataract, showing clouded portions of the anterior and posterior cortical regions. This dog inherited some visual impairment.*

*"Although this was a devastating experience,*

early onset include Alaskan Malamutes, Samoyeds, and Siberian Huskies.

### PRA/PRD

Discovered in European Gordon Setters in the early 1900s, progressive retinal atrophy/progressive retinal degeneration (PRA/PRD) is a catchall term for at least seven inherited bilateral, blinding retinal disorders. It is the canine equivalent of retinitis pigmentosa.

As a disease of the retina, PRA/PRD affects the photoreceptor cells inside the back of the eye, which comprise rods (vision in dim light) and cones (color, and vision in bright light). These cells absorb light that enters through the lens and convert it into electrical impulses. The

optic nerve transmits these signals to the brain, which translates them into what we perceive as sight. In most PRA/PRD cases the retinal cells deteriorate, starting with the rods and progressing to the cones.

Researchers have identified the genes responsible for what is now called “prcd”—progressive rod-cone degeneration—the major form of PRA/PRD found primarily in retrievers, spaniels, Portuguese Water Dogs, and Poodles.

*Prcd* can be “early onset,” where symptoms occur at 6 months of age, or “late onset,” which appears between 3 to 6 years. Both of these are referred to as “generalized” prcd PRA/PRD and inevitably lead to blindness. Gradual loss of sight in low light is the hallmark symptom of the disease but, on physical exam, the eyes may appear normal until night vision is severely compromised.

For years, the discovery of PRA in breeding lines struck dread into the hearts of breeders. But the pioneering work of researchers at Cornell University’s James A. Baker Institute for Animal Health, and the Fred Hutchinson Cancer Research Center in Seattle, has resulted in a DNA test that identifies, with close to 100 percent certainty, whether a dog is genetically free of the gene (Pattern A), a carrier (Pattern B), or one who will become affected by the disease (Pattern C). Carried out by Optigen Laboratories, of Ithaca, New York, results are disseminated based on breed-club decisions. There is no centralized registry.

Unfortunately, some of the earlier tests had a high false-positive rate, which contributed to the stigma.

Gerri Kennedy-Youngblood, of New Fairfield, Connecticut, endured four years of anguish believing that her stud dog Labrador Retriever, Captain, was affected and had passed on the gene to his offspring. “Despite CERFing clear through his 12th birthday, he was classified as a C when the PRA test first came out in 1999. I was devastated. He was the love of my life and figured heavily in my own pedigrees.”

Kennedy-Youngblood’s experience is not unique,

**With glaucoma, early diagnosis is critical to prolonging sight.**

CHINESE SHAR PEI/ISABELLE FRANCAIS FOR AKC



will not produce a PRA-affected dog.”

Gustavo Aguirre, VMD, one of the original researchers and a co-founder of Optigen, concurs. “You’re not breeding test results,” he says, “you’re breeding dogs. You want to keep your genetic diversity; if you exclude all carriers, you restrict your population too much and can come up with new diseases.”

Frances Paulen, DVM, a general practitioner, president of the Newtown (Connecticut) All Breed Kennel Club, and breeder of German Shorthaired Pointers and English Cocker Spaniels, says that PRA has been a sticky subject for too long. “There is still too much resistance and ignorance about PRA and the Optigen test. People are afraid to be ‘outed,’ but breeders didn’t create the disease—it’s genetic. Breeders are only responsible for creating it if they knowingly perpetuate it, and there is no excuse for not testing when a test is available.”

According to author Caroline Levin, the “silver

*the dogs are able to lead relatively normal lives.”*

but now has little chance of being repeated. In 2003, intensive further research brought the much-heralded second generation of tests for Labrador Retrievers and English Cocker Spaniels, adding the numeral 1 to the A, B, or C designation. Captain was reclassified as an A1 in July 2003, and Kennedy-Youngblood was “ecstatic.”

The incidence of false positives for carriers and affecteds is now significantly reduced. Kennedy-Youngblood is sanguine about her experience, and champions the test. “As a chemist and research scientist, I know that no test can be 100 percent accurate. But the improved marker test reduces PRA to the level of another ‘fault.’ You can confidently breed a B1 or a C1 to an A1 and be certain, to within a 0.5 percent chance, that you

lining” of PRA is that it happens gradually, does not cause pain, and dogs adjust readily.

Blind since age 7, Nina Skorus-Neely’s English Cocker Spaniel Roxy is a prime example. “At 9½, she is a superb dog. Blindness is not as bad as I thought it would be for her—she can sense the tennis ball, follow the other dogs onto the agility equipment, and retain her matriarch status.” Skorus-Neely is a vocal advocate for an open registry and heralds the Optigen test as a “powerful tool” to assist breeders in decision-making. 🐾

*Susan Lennon is a freelance writer who lives in Connecticut with her husband, two elderly yellow Labrador Retrievers, and one bossy black cat.*