RECOGNIZING MERLE
Newly Approved Coat Color Brings Change
It was one small step for an 11-month-old female Great Dane called “Bellini” (CH Dulce Put a Cork In It) when she stepped into the ring at the Great Dane Club of Maryland Specialty Show on April 13, 2019, but a giant step for Great Danes.

Bellini, bred and owned by Dr. Tracy Powell of Dulce Great Danes in East Windsor, Connecticut, became the first merle Great Dane in history to earn a conformation champion title. The newly approved coat color for Great Danes — merle Great Danes became eligible to enter American Kennel Club shows on Jan. 1, 2019 — signals a change in long-held beliefs about genetics. Shaping this new era is DNA testing, which has helped to provide definitive answers about recessives behind a dog and guidelines to aid breeders’ decision-making.

Merle has always been a Great Dane color but never an approved one. Throughout the past century merle has been a disqualification, meaning that regardless how...
elegant or regal an individual dog may have been, a merle Great Dane would be dismissed simply on the basis of its coat color. Even more problematic, breeding two merles together had a high probability of producing double merles, mostly white dogs with congenital deafness or visual impairments. Despite breeders’ best efforts to eradicate merles from the breed, the coat color continued to crop up.

Breeders disagreed about why merle remained in the gene pool. Some speculated that merle and harlequin were somehow related because harlequin Danes often produced merles. One of the most influential harlequin breeders was the late Laura Kiaulenas of BMW Great Danes in Farmingville, New York, who believed that harlequin resulted from the interaction of genes for black, merle and white. She was partly correct.

Geneticist Neil O’Sullivan, PhD, a Great Dane breeder working in the Department of Agricultural Zoology and Genetics at the University College of Dublin, Ireland, in the 1980s, was determined to find out. He began collecting breeding records from Irish and British breeders, amassing a large sample size of a variety of coat color matings.

Meanwhile, D. Phillip Sponenberg, DVM, PhD, professor of pathology and genetics at Virginia-Maryland College of Veterinary Medicine, began similar research in North America. In an article published in the Journal of Heredity in 1985, Dr. Sponenberg supported the theory that harlequin is a modification of merle.

Analysis of data collected by Dr. O’Sullivan and his team confirmed the hypothesis that harlequin was a modification of merle. This research was published in January 1988 in the journal Genetica. “In a U.K. litter, a harlequin Dane was produced from a merle x blue breeding. This was something the theory supported, and now we had an example in the field,” Dr. O’Sullivan says.

Most harlequin breeders accepted that without merle there could be no harlequin, but doubters remained. It would be decades before DNA technology could definitively explain the relationship. The opportunity came when a research team at Texas A&M University, led by the late Keith Murphy, DVM, PhD, professor of genetics, and Leigh Anne Clark, PhD, research assistant professor, identified the merle gene and then proposed a follow-up study to find the harlequin gene. The Great Dane Club of America (GDCA) was eager to support the research.

The project moved to Clemson University, following Dr. Murphy, who had become professor of genetics and head of the Clemson University Genomics Institute, and Dr. Clark, now associate professor of genetics. They located the harlequin gene and described how merle and harlequin interacted.

“By itself the Mm genotype causes a more colorful pattern, termed merle, characterized by dark spots on a dilute background,” explains Dr. Clark. “H is a dominant modifier of M that removes the dilute pigment and increases the size of the fully pigmented regions. H has no visible phenotypic effect in mm (wild-type) dogs; therefore, merle is the only phenotype that unequivocally indicates an hh genotype. The base coat color of harlequin and merle dogs is controlled by separate loci.”

The research was published in 2011 in Genomics. “We genotyped 270 Great Danes in our study and never observed H in homozygosity, which is consistent with Dr. O’Sullivan’s work,” Dr. Clark says.

This also explained why harlequin-to-harlequin matings seem to produce small litters. “Because
harlequin-to-harlequin matings are not associated with stillbirths, HH likely causes embryonic death early during pregnancy,” says Dr. Clark. “This is also why harlequins are not true breeding and almost always produce merle offspring.”

This research removed any lingering doubts about whether merle was essential to harlequin. It also spurred harlequin breeders into action. Breeders had already successfully petitioned GDCA to allow one color disqualification: mantle. Like merle, mantle is an integral part of the harlequin recipe.

Breeding merles or harlequins to mantles creates a high proportion of show-marked harlequins, which are required to have white trim in the Irish pattern on the collar, blaze and tail tip. Conversely, blacks produced from show-marked harlequins tend to have too much white trim to be shown. In recognition of this essential relationship of harlequin and mantle, GDCA added black mantle as an approved color in 2000.

Many breeders were keen to do the same with merle — especially since it was impossible to breed a harlequin without a great probability of producing merles. This put the harlequin breeder at a huge disadvantage as a large number of offspring from harlequin parents couldn’t be shown no matter how good they were otherwise.

Cindy Harwin, of Los Gatos, California, who has been breeding harlequin Great Danes under the Payaso prefix for 29 years, favored recognizing merles. “The reason we felt that merles needed to be recognized is that all of us who breed harlequins use them and produce them in our breeding programs,” she says. “Any dog that you use in your breeding program, you should be able to show. Because merles couldn’t be shown, many went to pet homes where, quite unintentionally, they had become considered an exotic color.”

Many breeders shared her opinion. In a 2015 poll of GDCA members, the membership voted by a large margin to allow merle as a recognized color. The standard was changed, and as of Jan. 1, 2019, merle Great Danes could compete in AKC show rings.

This brings us back to Bellini, who finished quickly with three majors. Dr. Powell, who handled Bellini to all but two wins, says, “It is an honor to be the breeder and owner of the first merle Great Dane AKC champion. The only merle puppy in a litter of 14 out of a harlequin dam and mantle sire, Bellini has always been a standout. No matter what color she is, she is an excellent example of the breed both in conformation and temperament.”

COLOR CODE CHANGES

Subsequently, other changes are coming. The Great Dane standard has always listed very explicitly what color combinations are allowed. Historically, the tight list of approved colors posed challenges, as interbreeding correct colors often produced disqualified color and pattern combinations such as harlebrin, tricolors, merlequin, and fawn mantle.

Early breeders found that by breeding only within certain color families — fawn/brindle, black/blue (solid-no white), and
The merle coat color gene comes from an extra bit of genetic material inserted into a pigment gene called \( S/LV \). All dogs carry the \( S/LV \) gene, which is responsible for producing a fibrous matrix within pigment cells that essentially holds pigment granules in place. If one \( S/LV \) allele has extra genetic material, the fibrous matrix is incomplete and pigment granules escape, leaving a faded coat color.

However, the genetic insertion is not very stable and tends to shrink. In some embryonic cells it shrinks to the point of being nearly normal, and the matrix these cells produce is almost complete. During development, cells derived from these near-normal embryonic cells give rise to patches of fully pigmented coat. Thus, merles are a mosaic of cells derived from cells with leaky matrices and normal matrices.

Recent research at Clemson University published in 2018 in *Mobile DNA* has discovered that merle coat color genetics are more complicated than previously thought. It turns out that the length of one part of the insertion determines the type of merle a dog will have. For example, dogs with an insertion of 25 to 44 base pairs in this area are cryptic merles with no obvious sign of merle. During development, cells derived from these near-normal embryonic cells give rise to patches of fully pigmented coat. Thus, merles are a mosaic of cells derived from cells with leaky matrixes and normal matrixes.

According to co-author Leigh Anne Clark, PhD, associate professor of genetics at Clemson University, most harlequin Great Danes have normal merle insertion lengths combined with one copy of \( H \) that together cause their harlequin appearance, and a few have the harlequin merle mutation.

“In our original harlequin study, there were two harlequin Great Danes that did not have the \( PSMB7 [H] \) variant,” she says. “We have since gone back to look at the insertion lengths in these dogs and found that one dog was 81 and the other 82. It turns out they were harlequin merles. Thus, in Great Danes, there are two ways to be harlequin.”

Breeders try to avoid producing double merles (\( MM \)) because is tied to a risk of congenital deafness and visual defects, the latter most often being a condition called microphthalmia, an abnormally small eye due to a malformation of the globe of the eye. But not all double merles are adversely affected, and most live full lives.

Payaso Great Dane breeder Cindy Harwin of Los Gatos, California, says she has owned three double merles — all were happy, well-adjusted dogs. “One was deaf, but you couldn’t tell by the way she acted,” she says.”These days fewer double merles are produced, and those that are tend to find a ready home.”

Marc Sayer, of Oakridge, Oregon, founder of Deaf Dane Rescue Inc., an organization devoted to double merle Dane rescue that takes in 10 to 20 dogs a year, says, “When it comes to disabilities, the rule of thumb is the less color, the more issues, but it’s not hard and fast.

“About 100 percent of our double merles have had eye issues. Generally the pupil is misshapen and/or off center, and often there is some level of microphthalmia. Most have star pupils to some degree,” he says. “If their eyes are different colors — one brown and one blue — the issues are limited to the blue eye, which isn’t really blue, rather, it lacks pigment so it appears blue. These problems affect almost all double merles, though most see well enough to have functional vision.”

A beautiful merle Great Dane, Bellini has a pale gray coat with black torn patches and a mantle pattern that includes a white muzzle, partial white neck, white chest, white on the forelegs and hind legs, and a white-tipped merle tail.
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DNA TESTING PROVIDES A RELIABLE COLOR TOOLKIT

Breeding within color families used to be the only way Great Dane breeders could control or predict the recessive color genes a dog carried. Now, DNA tests for coat color genes are available to help breeders. Here are examples of what you can expect to learn from coats color testing at the University of California-Davis Veterinary Genetics Laboratory.

- Dominant black versus recessive fawn (No test yet exists for brindle, which is at this same locus)
- Dominant mask versus recessive no mask
- Dominant black versus recessive blue
- Dominant black versus recessive brown
- Recessive spotted (No test yet exists for dominant solid or intermediate Irish-marked at this same locus, and technically a combination of solid and parti genes can produce Irish-marked)
- Dominant merle versus recessive non-merle
- Dominant harlequin versus recessive non-harlequin

Bellini, center on top, was the only merle Great Dane in her litter of 14 puppies.

accidental fawn-to-black mating. CH Honey Hollow Stormi Rudio (“Stormi”), the No. 1 Working dog in the country from 1957 to 1959, won eight Bests in Show and the 1959 Westminster Working Group.

Basquette followed with more color crosses, arousing the ire of fellow breeders while setting an unprecedented show record with her Danes taking 125 Bests in Show and 500 Group Firsts. Crossing color families enabled Basquette to introduce qualities from one color family to another, but at the expense of producing more puppies of disqualified colors.

She wasn’t the only one. J. Council Parker (Jocopa), who later became a respected judge, was first a renegade in the Great Dane world by using fawns and brindles to produce outstanding blacks. Others followed suit in an effort to bring traits from one color family into another.

“I know the motivation of these breeders, as I had close friendships with each of them,” recalls Dr. O’Sullivan, now vice president of GDCA. “They each felt trapped in a narrow gene pool (blacks and blues) and wanted access to dogs of superior quality that were outside their gene pool (brindles and fawns).”

The upside was a surge in quality, especially in the families that benefitted from the higher quality of the fawns and brindles. The downside was a cornucopia of non-allowed colors gracing the sofas of many a home. Stormi’s success, along with the increasingly visible color crossing,
brought attention to the Great Dane Color Code wars, with some breeders flaunting the code and others vilifying the scofflaws. The Color Code remained, and though the GDCA’s Code of Ethics demanded adherence to it, for many it was more of a guideline than rule.

Things change. That includes the Color Code, which has been taken off the GDCA website as it undergoes extensive revision. It will return, but with modifications taking into account advances in DNA color testing and inclusion of merle as an accepted color. Instead of DNA testing, the earlier Color Code relied on dogs being clear of certain colors for four generations to lessen the chance of them being carriers of unapproved colors. DNA testing provides clear understanding about the genes a dog carries.

Dr. O’Sullivan, who chairs the committee developing the updated GDCA Color Code, says, “The new color code will advise breeders how to get the colors they desire and avoid producing non-recognized colors. For example, in the black and blue colors, one should avoid breeding carriers of fawn or brindle together and at least one parent should be homozygous for dominant black allele (KK genotype). Among brindles, fawns and blacks, breeders should ensure at least one parent doesn’t carry a blue recessive allele as two blue carriers (Dd genotype) mated together results in blue fawn or blue brindle puppies.”

The former Color Code, developed before research showed that all harlequins have at least one merle allele, allowed breeding harlequin to harlequin. In such matings, on average one-quarter of the offspring are double merles, which have a high incidence of ocular problems and deafness. In one study published in 2009 in the Journal of Veterinary Internal Medicine, 25 percent of double merles were either unilaterally (10 percent) or bilaterally (15 percent) deaf.

“In some countries, like Germany, it’s illegal to breed two merles or harlequins together,” says Harwin, a member of the GDCA board of directors who is involved with developing the new Color Code. “In the U.S., we can’t set rules about what you can’t breed, so our new Color Code aims to be educational and give probabilities about what color combinations may produce non-standard colors.”

Dr. O’Sullivan says the new Color Code along with DNA testing are tools to help breeders. “Breeders will be able to pursue breeding better Great Danes without concerns over random color events resulting in their best puppies not being a recognized color or, worse, having a serious health defect such as deafness and/or blindness.”

Purina appreciates the support of the Great Dane Club of America, particularly Neil O’Sullivan, PhD, vice president and co-chair of the GDCA Health and Research Committee, in helping to identify topics for the Great Dane Update.
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*Data was collected by Relevance Research via an online survey from August 15-19, 2018. A total of 626 nationally-representative dog owners qualified and completed the survey. Qualified participants were men and women age 18 and older, owned one or more adult dogs, were household members most responsible for taking the dog(s) to a veterinarian, and had taken the dog(s) to a veterinarian in the past 12 months.

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