

Three Fatal Genetic Diseases in Show Greyhounds

by Patricia Gail Burnham

In the last decade, three fatal genetic diseases have been diagnosed in show Greyhounds. The first of these is Sub Aortic Stenosis (SAS), which is the most common congenital heart defect in dogs. The second is dilated cardiomyopathy which is an acquired heart disease with a genetic predisposition (it is quite common in Doberman Pinschers). The third is Osteogenesis Imperfecta (OI), which is a disorder of growing bones that generally shows up in puppies. Only mildly affected OI dogs will ever live to reach adulthood.

SUB VALVULAR AORTIC STENOSIS (SAS)

For years I have heard reports of Greyhounds in the prime of life that drop dead without warning. These losses occur both in this country and overseas. What can kill a dog without warning? Stroke (blood clots in the brain), aneurysms (weak spots on the blood vessels that rupture) and heart problems are the most likely culprits in sudden death. One of the heart problems that exist in Greyhounds and that can cause sudden death is Sub Valvular Aortic Stenosis (SAS).

SAS is caused by a narrowing of the aorta beneath the valve leading out of the heart. That is why it is sometimes called Sub-Valvular Aortic Stenosis. Sub Valvular means underneath the valve. Stenosis means narrowing, so Aortic Stenosis means a narrowing of the Aorta.

SAS is inherited as a polygenic dominant trait with variable expression. The variable expression means that an affected dog can have slight or severe narrowing of the aorta. Slight narrowing may cause mild heart murmurs and the dog can live a normal life span. Extreme

narrowing of the aorta causes sudden death in adult dogs. Prior to the fatal attack it may cause exercise intolerance, weakness and fainting spells. Or there may be no warning at all prior to death. SAS is hard to diagnose prior to postmortem although a murmur may be detected by stethoscope and a cardiac ultrasound can also help with diagnosis. The problem with the murmur is that there are lots of heart murmurs with other causes than SAS. How do I know that SAS occurs in show Greyhounds?

There is a family line of Greyhounds in which four half siblings and one double grandson have died unexpectedly as young adults. The first was found dead in his crate at a dog show. The second died while sleeping next to her owner. When we got up to dogs number four and five the owner had post mortems done and the diagnosis was SAS. As she said to me, "Greyhounds shouldn't faint." Their breeder had told her that their fainting wasn't serious.

When the aorta is narrowed, the heart has to work harder to pump blood through the aorta. As the diameter of the aorta decreases the speed of the blood increases and the blood pressure increases in an attempt to force blood through the narrowed opening. The heart beats harder to push blood against the resistance and because the heart is beating harder and faster it tires easily and may have abnormal heartbeats. A string of these abnormal heartbeats together may cause fainting episodes. Enough of them can cause a heart attack and death without warning.

Bouvier fanciers are trying to identify genetic markers for the genes that cause SAS in their breed and they have a good web

site under the subject Sub Aortic Stenosis Project. What they are trying to do is gain the ability to identify dogs that don't carry the genes for SAS to enable them to eradicate the condition from their breed in the future. The problem is not the dogs that drop dead before they have a chance to be bred. The problem is all the dogs who are affected but are not affected severely enough to remove themselves from the gene pool by dying before they are bred. These are the dogs that spread the genes for SAS.

Since SAS is inherited as a polygenic dominant, if a dog with the trait is bred to a non-affected dog, half the puppies should be affected. They are not carriers, as they would be with recessive genes, they are actually affected, but they may be affected sub-clinically.

From the affected patterns in Greyhounds, it appears that if you double up on an affected ancestor, the degree of SAS becomes more severe. Three of the severely affected dogs were the result of half brother-sister or father-daughter breedings that increased the likelihood of the offspring being homozygous for SAS.

One nice thing about inbreeding is that it does reveal both recessive and polygenic dominant faults by intensifying them. But the real problem is that the siblings of the affected dogs have a fifty percent chance of also being affected. And these dogs may not be affected enough to show symptoms, which means that they might be bred.

My ophthalmologist has always owned male Bouviers. This week he told me about buying a new puppy and having to get a bitch. (He was in shock about how different the girl's personalities are.) The breeder had taken the

whole litter to a cardiologist to be screened for SAS and both males had been affected. So she would only sell him the bitch that had not shown a heart murmur. I found this interesting because most of the literature on SAS says that it is hard to diagnose without a post mortem. Perhaps the breeder was just refusing to sell any puppy with a heart murmur. (The increased speed of the blood flowing through the SAS restriction can be heard as a heart murmur.) But how could one tell an SAS heart murmur from the other kinds of murmur.

My canine cardiologist says that he can often tell serious SAS from the other forms of heart murmurs by an echocardiogram, but only if it is a grade two or higher murmur. Murmurs are graded from one to six by severity, with six being the most severe and one the least. Heart murmurs aren't a great indication of SAS, because heart murmurs have many different causes and most of them are benign so they cause no health problems. In human children, 60% of their heart murmurs are benign.

I suspect that Greyhound puppies get diagnosed with heart murmurs more often than other breeds of dog because their hearts are larger and pump more blood than the hearts of dogs that weren't developed for racing. My regular vet has twice referred me to his cardiologist because Kira's heart sounds odd to him. Each time the cardiologist has given her a clean bill of health. I am willing to concede that her heart sounds different than the hearts of other dogs that my internist listens to. But that doesn't mean that her heart is not healthy.

Dogs aren't certified for cardiac health until after they are a year old, but heart problems can be diagnosed at younger ages. However, at any age, the mild cases may be missed on diagnosis.

That is why the Bouvier fanciers want a genetic test for the gene. It will enable them to tell the mildly affected dogs from the dogs that don't carry the gene at all.

CARDIOMEGALY AND CARDIOMYOPATHY

In a separate family group, a sire and his daughter both died suddenly as young adults. The sire collapsed and died while barking at a neighbor's dog, and the daughter collapsed and died while in the house with her owner. A post mortem on the daughter identified her heart problem as cardiomegaly. Cardiomegaly simply means enlargement of the heart. It is often a response to bad heart valves or weak heart muscle. When the valves don't seal completely, then blood flows both forward and backward when the heart contracts. When the heart muscle itself is weak, then the heart is pumping inadequate blood with each contraction. In order to compensate for the inadequate flow, the heart enlarges to try to pump more blood to the body. The autopsy identified cardiomegaly with ventricular hypertrophy, and mild valvular endocardiosis.

Diseases of the heart can be subdivided into cardiac malformations (birth defects of the heart or major blood vessels) and acquired heart disease. Malformations are present at birth and include the sub aortic stenosis (SAS) discussed above. Acquired heart diseases are not present at birth but are acquired later. Many acquired diseases have a genetic basis, but the clinical condition is not evident until the dog grows. Cardiomegaly is one. Cardiomyopathy is another. Cardiomyopathy is a hereditary, acquired disease that affects the heart muscle. The effect of cardiomyopathy is reduced heart muscle contraction,

which can lead to heart failure. The second effect is electrical instability of the heart (arrhythmia) that leads to a heart rhythm that is too fast, too slow or erratic. These arrhythmias can cause fainting or sudden cardiac death. Heart enlargement is often a by-product of cardiomyopathy as the heart tries to compensate for its decreased pumping ability.

For our foreign readers who are congratulating themselves on not having heart problems, the sire of the SAS family and the sire of the father daughter pair that died of heart problems were both imported dogs. One was from Scandinavia, the other from England.

OSTEOGENESIS IMPERFECTA (OI)

Because OI affects both dogs and people, most of the good sources for information about it are the human ones. The OI Foundation web site is particularly informative.

OI is a collagen deficiency disease that affects the bones. Structurally, bones are made up of calcium compounds and collagen. (I am ignoring the fatty marrow where the blood cells are produced.) It is the collagen in bones that makes them somewhat flexible. Think of the difference between raw chicken bones and cooked ones. Raw bones are flexible. When you cook them, you cook the collagen out of them (which forms a nifty broth and thickens when it is cooled). But the once the collagen has been removed from bones, they splinter instead of bending. OI is a disease where a puppy (or person) either has less collagen than normal or that collagen is abnormal. This causes weak bones that fracture easily.

Some parents with OI children have been accused of child abuse before their children were properly diagnosed. In humans, OI is generally inherited as a dominant genetic defect. Recessive

inheritance is rare. When OI occurs with no family history, it is considered to have been a spontaneous mutation but it could also be the result of recessive genes inherited from each parent. As with SAS, the OI gene can be expressed mildly or severely.

In people, OI is divided into four recognized types. Type I is the most common and mildest form. In Type I the collagen structure is normal but the amount of collagen is less than normal. Type I is the mildest form of OI. In types II, III and IV, which are all more severe, the collagen is improperly formed.

People have one advantage over dogs. Nobody inbreeds people, so the recessive or dominant genes have little chance of matching up with another of the same gene. When we line breed and inbreed our Greyhounds, we vastly increase the chances of matching up like genes. If a puppy gets a mildly affected OI gene from each parent, the result may be a puppy with severe OI. These are usually the only puppies in which OI is identified. They suffer multiple fractures early in life, often before they are weaned. I only know of one puppy that managed to reach a home before being diagnosed with OI. Usually they get put down before they are weaned. If OI in dogs is indeed inherited as a dominant trait with variable expression then, when an OI puppy appears, at least one parent must have a mild form of OI. It is possible that both parents have the genes for a mild expression of OI, and the doubling up of the genes is what caused the increased severity in the puppy.

The two litters that I know of with OI puppies were only related distantly. And they were not related at all to the SAS family above. OI seems to be a more American problem, since the pedigrees of both litters go back for generations before reaching imported dogs.

Bitches have been stepping on and squashing puppies for as long as there have been puppies, but the broken bones in OI puppies tend to be multiple and repetitive. And the bone breaks are so painful that the kindest treatment for OI puppies is euthanasia. Human cases don't have that option, and there is a lot of information on the web sites devoted to the care of children with OI.

NOTE: The prolific sire, Foxden Flamingo, broke three legs and his jaw. He was born in 1943 and his poor bone quality has been ascribed to his poor

nutrition while growing up in England during World War II. But his brittle bones could easily have been a moderate expression of OI.

The above diseases, along with liver shunts and demyelination of the spinal cord, are some of the genetic issues beginning to emerge within our breed. ■

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- BOB** 73 CH JONZ CONSTANT CRAVING. HM842616/01. 10/6/1996. Dog. Breeder: John R Palmer. By Ch Jonz Obsession x Ch Jonz Unchained Melody. Owner: Dana K Olson & John Palmer. 22555 County Rd 5, Elbert, CO 80106.
- AOM** 75 CH HYPERION CHRISTMAS PRESENT. HM931843/02. 11/17/2000. Dog. Breeder: Dana K Olson. By Ch Gayside Christmas Past x Ch Legacy's Incredible Journey. Owner: D Olson & B Hargreaves-Savage & I Savage & J Palmer, 22555 County Rd 5, Elbert CO, 80106.
- 78 CH SOUTHWESTERNS SIERRA'S INCA. HM521036/01. 5/16/1994. Bitch. Breeder: R Owens & G Davis & Anne Tater. By Ch Alzanna's Dallas x Ch Another Episode Skylite. Owner: Dr S Hempel & G Davis & R Owen & T Hendricks, 6318 Ben, Sachse TX, 75048.
- 80 CH WILLOWMOOR TWICE TOLD LEGEND. HM747037/04. 10/9/1997. Bitch. Breeder: Patti Clark & June Matarazzo. By Ch Jonz Devil Made Me Do It x Ch Willowmoor Divine Madness. Owner: Pamela Noll, 11802 Pinto Cir, Buda TX, 78610.
- 82 CH GREYCOTT HAPPY TIMES. HM756645/03. 10/10/1997. Bitch. Breeder: Owners. By Ch Shazam's The Journey Begins x Ch Heirloom Lovin Life. Owner: Scott Thompson & Bruce Clark, 5559 S Washington, Littleton CO, 80121.
- AOM** 84 CH SIERRA'S MIATA ROCKET. HM778339/03. 6/16/1998. Bitch. Breeder: Dr S Hempel & B Hempel & R Owens & G Davis. By Ch Rockets Oracle x Ch Southwestern Sierra Inca. Owner: Dr Shelley & Bill Hempel, 6318 Ben Rd, Sachse TX 75048.
- 86 CH BRASSPUR OAKLYN BLUE MIST. HM818013/05. 1/19/1999. Bitch. Breeder Rick Metts. By Ch Jonz Huzzah Heavenlybody x Ch Legacy's Incredible Journey. Owner: Karen Lynn Ahlquist, 6408 Ben Rd, Sachse TX 75048
- BOB** 88 CH GREYCOTT OH FOOLISH MOON. HM893720/05. 4/1/2000. Bitch. Breeder: Scott Thompson & Bruce Clark & R Unger. By Ch Jonz Huzzah Heavenly Body x Ch Heirloom Lovin Life. Owner: Deborah Bahm & Lori Wilson & Scott Thompson, 23456 Chat Blanc Ln, Bush LA (Lori Wilson, Agent) ■

SNUFFY'S MAGIC LIVER TREATS

1 lb raw liver
3/4 cup grated cheese
1/2 cup pureed carrots
2 eggs
4 tablespoons butter, melted
4 cloves of garlic
3/4 cup finely crushed graham crackers
1/3 cup rye or whole wheat flour
1/2 cup (or more) of corn meal

Put the liver and one egg in a blender or food processor; puree until liquid. Place carrots, the other egg, the melted butter and the garlic in the blender and also liquify. Add the carrot mixture to the liver mixture. Add the cheese, graham crackers and flour. Mix together. Add the corn meal; mix; consistency should be very thick—just like wet cement. If needed, add more corn meal until you get wet cement. Spread on a lightly greased piece of tin foil on a cookie sheet. Bake at 350 degrees for 30 minutes. Let cool completely and then cut up into cookies. Will keep for several weeks in the fridge and freezes well.

—Eric and Nancy Liebes