

Neck Pain

By Dr. J.E. Dillberger

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The Deerhound Health Survey revealed that neck pain was one of the top dozen health problems in our breed. According to survey data, the incidence of neck pain is just under 6%, meaning that about 1 in 18 dogs will experience an episode of neck pain during its life.

These data did not surprise me. I already suspected that neck pain was common in our breed based on how often I'd talked with fellow Deerhounds about the problem, which is common enough to have been dubbed "deerhound neck" by some fanciers. Also, two of my own hounds have had neck pain, so I've experienced the problem firsthand.

Most Deerhounds whose dogs experienced neck pain have sought veterinary care, and these folks have received various explanations for what caused the pain. Often Deerhounds have associated an episode of neck pain with some preceding event, and presumed that the event caused the pain. No clear single cause for this problem has emerged.

Let me say up front that I do not know if all Deerhounds with neck pain suffer from the same underlying problem, nor that neck pain results from a single cause instead of from the interaction of multiple predisposing factors. I do know that the similarity in the symptoms, clinical course, and response to treatment of neck pain in different hounds over many years suggests that a neck pain syndrome exists in our breed. My own observations, discussions with other veterinarians, and a review of the medical literature have led me to a theory about what may underlie this neck pain syndrome.

In this article I'll describe the neck pain syndrome in our breed as I've distilled it from my own and others' experience. I'll present my own theory about what causes or contributes to this problem, laying out what evidence I have and pointing out where I'm only guessing. Finally, I'll include an interview with Purdue University's Dr. Larry Glickman about neck pain.

Deerhound Neck Pain Syndrome

A syndrome is a set of symptoms that occur together, in other words, a symptom complex. Symptoms of neck pain syndrome in Deerhounds include:

- ***Grunting, yelping, or crying out*** when laying down, getting up, shifting positions, stretching, or bending down to eat or drink. An affected dog may be reluctant to eat or drink at all, unless the bowl is elevated sufficiently.
- ***Reluctance to bend the neck.*** An affected dog often holds his neck rigidly extended straight in front, parallel to the ground. To avoid bending his neck sideways, a dog will follow a moving object with only his eyes or turn his whole body to keep the object in view, instead of turning his head to follow it. He'll back out of a tight spot, instead of bending his neck or body to turn around.
- ***Pain when the neck is bent.*** Sometimes any attempt to move the dog's head to the side or up and down elicits pain. Some dogs resist even having their head touched, their mouth opened, or their ears examined. In extreme cases, dogs will refuse to eat even from an elevated bowl because chewing is too painful.
- ***Fever.*** After a few episodes, I could detect this in my own dog without recourse to a thermometer, just by noting his bright red gums and hot ears.
- ***Episodic course,*** with alternating painful and normal periods. Pain and fever tend to come and go on their own, regardless of any treatment or the lack thereof.
- ***Young age.*** The typical neck pain sufferer is a younger hound. According to the Health Survey, the average age of the first pain episode is about 4 years old, and males are affected more often than females.

Because neck pain is episodic, it subsides on its own. If pain subsides while a dog is being treated in some way, then the treatment may be credited incorrectly with producing the relief. On the other hand, any symptomatic treatment that eases pain in general is likely to help dogs with neck pain syndrome, which is why pain relievers (such as aspirin, Advil, or Tylenol), massage, acupuncture, chiropractic adjustment, and certain herbal medicines make affected dogs more comfortable. However, as far as I can tell, none of these treatments has clearly ended an episode of neck pain or prevented its recurrence.

My Own Neck Pain Theory

When I observed the first episode of neck pain in one of my Deerhounds, I already was working as a toxicologic pathologist for an Indianapolis pharmaceutical company, helping develop new human and veterinary drugs by testing them in animals. Among my jobs was to examine microscopically tissues collected from Beagles that had been given promising new drugs to determine if the drugs caused any harmful effects that physicians and veterinarians should be aware of when they first tested the drugs in patients.

One compound that I studied looked potentially useful as a treatment for HIV infection (the virus that causes AIDS) in people, but it caused an inflammation of the blood vessels in dogs that received high doses. The general term for blood vessel inflammation is *vasculitis*; if the affected vessel is an artery, then the term *arteritis* may be used instead.

Vasculitis in Beagles was not new to me. I'd seen it occasionally in tissues from normal Beagles. So had other pathologists; in fact, vasculitis is considered a "background" pathologic change in the breed, meaning it shows up occasionally in dogs that are outwardly normal. However, the anti-viral compound that I was studying seemed to trigger vasculitis in Beagles. If such drugs existed, and they had proven safe in people, then I would feel more confident that my drug also would be safe.

My literature search turned up numerous articles describing drugs that caused vasculitis in Beagles, some of which had subsequently been tested in people and caused no harm, so I was reassured. But the search also turned up other articles whose titles and summaries caught my eye as a Deerhound owner. I want to share bits of five of them with you.

An Idiopathic Febrile Necrotizing Arteritis Syndrome in the Dog: Beagle Pain Syndrome (1989)

Idiopathic means of unknown cause, *febrile* means accompanied by a fever, and *necrotizing arteritis* means an arteritis in which the blood vessel wall is destroyed. The authors coined the term "Beagle Pain Syndrome" (BPS) to describe an illness they studied in 14 dogs. BPS was characterized by "intermittent pain, fever, neutrophilia (elevated white blood cell count), and arteritis." The authors described BPS as "a latent condition" that could be activated in predisposed dogs by experimental treatment. Typical clinical signs of BPS were "pain when the mouth is opened, grunting when lifted, and standing with an arched back and lowered head." In the authors'

experience, these signs usually waxed and waned, but could be persistent. Microscopic examination of tissues revealed widespread arteritis that often occurred in the cervical spinal cord and *meninges* (the membrane that surrounds the spinal cord), sometime accompanied by bleeding or blood clot formation. Over time and with repeated episodes, muscles in the temples and neck atrophied (wasted away), and dogs developed *amyloidosis*, a condition where abnormal protein material accumulates in multiple organs and impairs their function.

Systemic Necrotizing Vasculitis in Nine Young Beagles (1992).

The authors studied 9 dogs with a syndrome they called “juvenile polyarteritis syndrome” (JPS), characterized by fever and signs of pain in the neck area. Dogs with JPS had a hunched stance and were unwilling to move. Clinical signs resolved rapidly with high doses of corticosteroids, but in untreated dogs signs had a “remitting and relapsing course” that typically alternated 3 to 7 days of illness with 2 to 4 weeks of remission. Microscopic examination of tissues collected at necropsy revealed necrotizing arteritis that most frequently affected vessels in the cervical spinal cord.

Steroid-Responsive Meningitis-Arteritis in Dogs-Long-Term Study of 32 Cases (1994).

(*Meningitis* means inflammation of the meninges.) The authors separated their cases into two types: dogs that had a “typical form” with neck rigidity, pain, and fever, and dogs that had an “atypical form” with neurological deficits as well as neck pain and fever. All dogs responded to corticosteroid treatment initially, but the problem recurred. After long term steroid treatment, twelve dogs were “considered cured,” meaning that they remained pain free for several months, but thirteen dogs were euthanized because they remained in pain or had progressive neurological disease. Microscopic examination of tissues revealed arteritis of the meningeal arteries.

Pathologic Features of Naturally Occurring Juvenile Polyarteritis in Beagle Dogs (1995).

This paper describes in detail the microscopic features of polyarteritis in 18 dogs who had JPS. Arteries in the cervical spinal meninges were “consistently involved”, with “massive accumulations of inflammatory cells” that “often extended into adjacent tissues.” Sometimes blood vessels became obstructed by blood clots that formed at the site of inflammation. Eight dogs that had experienced repeated episodes went on to develop amyloidosis in liver, spleen, and kidneys.

Delving further, I found papers that described similar syndromes in non-Beagle dogs, although most of these papers reported only a single case and rarely included a definitive diagnosis. This is only natural, as the arteritis that is responsible for the clinical signs of illness in Beagles (and presumably other breeds) with this syndrome is detectable only at necropsy, when spinal cord tissue can be collected and examined microscopically.

Let me summarize what I had learned from reviewing the medical literature. A syndrome characterized by neck pain and fever occurs in many dog breeds, but is especially well described in Beagles. This syndrome has been called “canine pain syndrome,” “Beagle pain syndrome,” idiopathic polyarteritis,” and juvenile polyarteritis syndrome.” Affected dogs hold their neck rigid and are reluctant to move. If untreated, the signs of illness come and go in cycles. Treatment with corticosteroids eliminates symptoms, but they often recur when treatment stops. At least in Beagles, this syndrome usually occurs in young dogs (first episode before 3 years old). Microscopic examination of tissues from affected dogs reveals arteritis that can involve many tissues but almost always involves blood vessels in the cervical spinal cord and membranes that surround it. This arteritis typically is severe, with massive influx of inflammatory cells and vessel wall damage. Vessel wall damage may either cause a blood clot to form, which obstructs blood flow, or cause the vessel wall to rupture, which allows blood to escape into surrounding tissues. The severe inflammation accounts for the fever in affected dogs. Presumably the inflammation also accounts for the pain, either as a direct consequence (recall how painful a boil can be) or as a result of spinal nerve damage due to pressure from bleeding or lack of blood supply.

The clinical description of this syndrome in Beagles and other breeds certainly sounded very similar to what I had observed in my own Deerhounds, but I had no evidence to connect the two until last year. That’s when the testicles on one of my Deerhounds who suffered from episodic neck pain slowly shrank. One testicle became soft, while a firm round mass developed in the other. These testicular changes occurred gradually during a period when this hound was having neck pain episodes. I was worried that the mass might be a tumor, so I neutered him and examined the testes.

What I found in the testes was not a tumor, but arteritis! Besides active inflammation in several testicular arteries, I also found bleeding around

damaged vessels and blood clots within vessels. The testes had areas of necrosis (tissue death) due to lack of blood supply. On top of all that, I found scars from past episodes of vasculitis and necrosis as well. The mass that I'd feared was a tumor actually was a large lump of scar tissue.

Now I knew that my hound had experienced bouts of arteritis in his testes at the same time he experienced bouts of neck pain. Here was compelling circumstantial evidence that the neck pain syndrome in my Deerhound might be the same syndrome in other breeds and studied extensively in Beagles. And if that were true in my dog, I reasoned, then this same neck pain syndrome might account for at least some cases of neck pain in other Deerhounds, too.

Based on my theory, I was anxious to learn more about research on the neck pain syndrome in Beagles, and whether there was any way to distinguish the syndrome from other possible causes of neck pain. Ideally, I wanted to talk at length with someone involved in the published research studies. When I looked at the authors listed on key papers, I noticed a familiar name: Larry Glickman. I'd already talked with Dr. Glickman about his bloat research and on several other subjects, but had no idea he'd also been researching neck pain.

An Interview With Dr. Glickman

I arranged to interview Dr. Glickman at the 1997 National Specialty, which he attended in order to present his bloat research result. Excerpts from that interview follow:

Did you first get into the neck pain problem through working with Beagle colonies?

I got into it first as a consultant for Acme Breeders (fictitious name). I started with Acme in the latter part of 1978. I was at Cornell, on the faculty there, and got a call one day:

"Are you an epidemiologist?"

"Yes."

"Well, we don't know what an epidemiologist is, but someone suggested we call you."

They described how they were losing a third of their puppies to a diarrheal disease. They had no idea what it was. They'd had a parasitologist in, a

bacteriologist in-nothing seemed to help. That was the first reported outbreak of parvovirus. They went from virtually no mortality to 30-40%. When you have up to 7,000 dogs, that's a lot of diarrhea.

This turned out to be the first reported parvovirus outbreak. We not only identified the cause, but helped develop the first parvovirus vaccine, which eventually curtailed the outbreak.

So after that, they decided that an epidemiologist was a good consultant to have?

Yes. But to get back to neck pain...

I was walking through their kennel one day, and someone said, "You want to see a funny acting dog?" It was sort of standing in a corner with its head down. When I reached for it, it sort of turned to nip at me, which Acme Beagles never do. I've probably handled 150,000 dogs there in my life and been attacked once, and it was a bitch with puppies.

I asked them how often this occurred, and they replied "Oh, it's very sporadic, but we see it."

Then I went back to their records and tried to make heads or tails of it. Since then, we've instituted a monitoring program, looking at the frequency, pattern, genetics, etc.

Acme keeps good pedigree records?

Yes. Eight generations, all computerized. For any dog, I can calculate its coefficient of inbreeding and its relatedness to any other dog in the kennel.

The genetics of neck pain syndrome are not very obvious in that colony. It's clear it is genetic, and that a few sires stand out in the pedigrees of all these dogs, but it's not a simple pattern.

We started doing research on it. Acme would donate dogs. They wanted to be open about it. And it was a very low frequency problem. We actually did end up breeding two affecteds to each other, which is very hard to do, and we ended up with three out of five in the litter affected.

We were called by people at Harvard University in the Center for Blood Research, and they said, "Everything we've read about what you're doing sounds like this is a model of Kawasaki's disease in kids. Would you like to collaborate with us?" We worked with them for a few years. We tried to get an NIH grant on it, but couldn't sell it because of the very low frequency.

One of our pathologists is now sort of taking the lead. He probably autopsies about 20 of these dogs a year.

How did you link the vasculitis as a lesion to the clinical syndrome?

On the first few dogs we necropsied, we looked at the cervical spinal cord, including the blood vessels. What was striking was that not only the vessels in *that* area, but in some dogs the coronary vessels...

That's what I'd seen reported mostly in the literature, before your own papers, was coronary vasculitis.

That's true, but it was the dural blood vessels that really struck us. [Note: the dura is part of the meninges.] We've even had a few dogs that had mesenteric vessels involved, and bled out. And we saw the small vessels in the testicles affected. If you follow the dogs long enough through episodes, they'll almost all develop amyloidosis-widespread, including in the thyroid gland.

The natural history of the thing is interesting, too. If you do nothing, dogs go through exacerbations and remissions with cycles of variable lengths. We found in the laboratory, that if we have enough dogs we can synchronize their cycles with corticosteroids. If they have an episode and you blast them with corticosteroids, you can almost count 2-3 weeks and they'll have another episode.

Is this hard to diagnose from other neck and back pains? What are the distinguishing characteristics?

First of all, the age is rather typical. The range is broad, but typically the first episode occurs between four and eight months of age. We've seen it as early as two months, and occasionally in a 3- or 4-year old, but that's unusual. So, the age, coupled with fever of unknown origin, coupled with characteristic gait and neck held down.

We videotaped affected dogs, and as soon as we distribute or show the video, people say, "Oh yes, I saw that last year. I thought it was a disc problem," or "I thought it was meningitis. I treated it with antibiotics and it went away."

I'll say, "What else did you give it?"

And they'll say, "I gave it steroids, too."

In Deerhounds, neck pain often is attributed to trauma, because it usually occurs in puppies at an age where they're growing real fast, they're clumsy, and they're big. They all take multiple falls. That's just part of growing up in your first couple of years, so it's very easy to link neck pain to some episode like that.

Clearly there's a breed predilection because even in Beagles, it's a very sporadic disease. It's reported a lot in Beagles, but I don't think there's a breed predilection. I think the Scottish Deerhound, the PBGV...

What about the Irish Wolfhound? They're genetically related to the Scottish Deerhound.

I have never seen an affected Irish Wolfhound.

It wouldn't surprise me if there are multiple risk factors behind this syndrome. In a Beagle colony, the incidence may be lower than it might otherwise be, because they're confined dogs with a very defined environment, regular feeding, etc. Those dogs aren't exercised hard and they aren't getting traumatized.

Exactly. They have no other precipitating factors. Although, you talk about stress...this may be one disease where stress is helpful.

Every time Acme kennels has a Beagle that has this syndrome, they immediately ship it to us. We know that normally the cycle would go about 6 days if untreated. They usually ship on the first or second day. We almost never have a dog arrive with acute signs. We're pretty sure that's because the stress of shipping and the steroid release that accompanies it mimics treatment.

Is the lifespan of affected dogs shortened?

We've never followed them out. The longest we've ever kept a dog is about two years. They'll have repeated episodes, and seem to get longer between episodes with age. When you examine tissues from such dogs, they have all the classic chronic lesions.

Neck pain often shortens the life span of pet dogs because owners put them down; they can't live with repeated episodes of excruciating pain, with their dog screaming and unable to get up or lay down.

You can't touch these dogs!

This kind of episodic disease sets itself up for anecdotal treatments that "work," doesn't it?

Everything will "work!" That's another reason that NIH lost interest in this as a model for evaluating treatments for Kawasaki disease in kids—they realized that it's very hard to evaluate therapies in this model because of its exacerbations and remissions.

I have a hunch about one thing that may work that hasn't been tried, which may be curative. We've published a couple papers that you might have seen on the use of human intravenous gamma globulin to treat immune-mediated diseases. The idea came from what's done in kids with vasculitis, where for a long time physicians were using either aspirin or steroids, but found you got recurrences that were almost worse than the initial episodes before treatment.

For reasons I can't recall, physicians started using intravenous gamma globulin. It's really rather remarkable because one very, very high dose cures the problem in these kids. At the moment there's some controversy about whether the coronary artery disease that's seen early in life (in the '30's in some men) may be related to Kawasaki disease that was treated when they were young. Regardless, the clinical signs just ameliorated.

That's the reason I started using intravenous gamma globulin for a variety of immune-mediated diseases in dogs. Canine gamma globulin isn't available

commercially, so we use the human, which caused a lot of concern initially. But those concerns proved unfounded.

For example, in dogs with autoimmune hemolytic anemia, who have failed steroid treatment and have had tens of blood transfusions, about a third will be cured with a single gamma globulin infusion. Some may require two. It fails completely in about 50% of dogs.

Even though it's a human product, we've still had no side-effects with it. It's usually infused over 3 or 4 hours. During the infusion, we've had one or two dogs that spiked a fever. Stop the infusion and it goes away, and then you can start again.

Something even more remarkable: these dogs have almost no reticulocytes in their circulation. [Note: reticulocytes are immature red blood cells, which can be released from bone marrow early if the demand for red cells is great.] Their bone marrow just isn't putting out any reticulocytes. Within 20 minutes of starting the infusion, the reticulocyte count begins to rise. It's rather dramatic, and of course following behind it is the rise in the packed cell volume.

We've had very good success with that disease. We've had very good success with myelofibrosis and anemias--profound anemias. These successes have all been in pet dogs. We've now started using gamma globulin for thrombocytopenia, with the same results. So I think there's a reasonable probability that dogs with vasculitis-caused neck pain will respond to this type of intervention.

Advil brought more relief for my own dog with neck pain than did anything else.

It does in our dogs. The nonsteroidal antiinflammatory agents like Advil bring relief.

During my dog's last episode, you could run your hand down his back and feel the heat at about the fourth thoracic vertebra. It's easy in Deerhounds because they don't carry much body fat; there's just skin and a little lumbar muscle.

There's tremendous inflammation around the blood vessels, so I'm not surprised that you might feel heat.

That's true, but at the same time, it confused me. I thought, "Does this dog have an infected vertebra?" Because one of his testicles was soft by that point, I was worried about brucellosis.

We've seen lesions virtually any place, which means you can have pain virtually anywhere. One dog would not let you get near its face; it went absolutely crazy if you went near its mouth. We radiographed the dog's head, and it had a vasculitis in the vessel going into the skull. There was tremendous inflammation, with bone reaction around the vessel where it entered the bone. The dog must have been in excruciating pain.

I can see that an advantage of studying neck pain in Deerhounds or PBGVs is that you can get a handle on some other contributing factors. On the other hand, you won't get the kind of pedigree information that you get from Acme, and you're going to run into resistance among people admitting that they've had this problem or that they're aware of it. I've run into that. I'd like to think that they've never had it or seen it, but I think there's some denial going on. There's some understandable fear that the problem will be linked to a particular line.

If you want to investigate the problem in Deerhounds, then you ought to collect serum and DNA from affected dogs.

I think vasculitis clearly is an immune-mediated disease. The question is, "Is it an **auto**-immune disease?"

We'd like to get some serum from dogs with vasculitis, and I don't know how much it matters whether you get them during an acute episode or not. As a side benefit of other research, we now have ELISA assays to screen serum for about 14 antibodies (actually auto-antibodies).

We'd like to go back and screen all our vasculitis dogs, which are all Beagles with the exception of one or two, and then screen a couple serum samples from Deerhounds to see if we detect any auto-antibodies.

How much serum do you want, and do you just want it frozen?

Frozen serum would be fine. 1-2cc would be plenty.

You mention Breeds other than Beagles. Have you picked up much neck pain and vasculitis in other breeds?

Yes. As soon as we published our studies in Beagles, cases in other breeds started to surface. In fact, the Petit Basset Griffon Vendéen breed club will tell us soon if they'll fund us to work on their breed, because they're talking about 20-30%* being affected. They're having a real problem with it.

Let me summarize this long article as briefly as I can. Neck pain is a fairly common problem in Deerhounds. The clinical features of neck pain in some Deerhounds, including my own, resemble a neck pain syndrome that's been studied extensively in Beagles and reported occasionally in other breeds. Affected dogs have repeated episodes during which they hold their neck rigid, resist bending their neck cry out in pain when their neck is moved, and run a fever. Between episodes, dogs are normal.

In Beagles, the neck pain syndrome results from vasculitis (inflammation of the blood vessels) in and around the cervical spinal cord. This vasculitis often is widespread, occurring in many other organs. To my knowledge, no one ever has examined spinal cord tissue from a Deerhound with neck pain to look for vasculitis; however I have found ongoing vasculitis involving testicular blood vessels, as well as scarring left from previous vasculitis episodes, leading me to suspect strongly that the neck pain episodes in my dog were caused by vasculitis. Thus, I'm fairly sure that the neck pain syndrome described in Beagles occurs in Deerhounds, too and accounts for at least some of the neck pain reported in the Deerhound Health Survey.

There currently is no diagnostic test for vasculitis, but Dr. Glickman at Purdue University has an experimental assay that might be useful for screening serum samples from affected dogs (either during or between pain episodes). Once enough samples have been tested from Beagles and other breeds, Dr. Glickman can determine if the assay will become useful as a diagnostic test.

**Dr. Roughie regards the "20-30% being affected" statement as exaggerated.*

Dr. Glickman also suspects that commercially available human gamma globulin might be useful as a treatment for vasculitis-related neck pain, but he hasn't yet tried the treatment on any affected dogs. I intended to try this treatment on my own dog earlier this year, however, I couldn't locate a source of human gamma globulin, which, like other human blood-derived products, is in very short supply.

In closing I would urge Deerhounds whose dogs have had or now have neck pain episodes to consider sending a serum sample to Dr. Glickman at Purdue University with a letter describing the dog's health problem and requesting that the serum be tested in his experimental assay. Furthermore, I would urge Deerhounds whose dogs have suffered neck pain episodes during life to have a necropsy performed when the dog dies, and to provide a copy of this article to the person who will perform the necropsy, with explicit instructions to examine tissue from the cervical spinal cord. Only in these ways can we discover how much of the neck pain in our breed is due to a problem shared by many breeds.