Canine Caval Syndrome Series

PART 2: A PRACTICAL APPROACH TO DIAGNOSING CAVAL SYNDROME

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Not long ago, heartworm disease (HWD) was considered a problem of the South—it was endemic in the Southeast, Gulf Coast, and Mississippi Delta regions of the United States. Today, we find heartworms not only present in, but also transmitted in, every state in the continental U.S. as well as Hawaii (Figure 1).1

According to the American Heartworm Society, over one million dogs in the U.S. are currently infected with *Dirofilaria immitis* (heartworms). Although most infected pets are clinically normal, it has become increasingly important that veterinarians everywhere recognize the clinical signs of illness.

A small percentage of dogs infected with heartworms develop caval syndrome (CS), an extremely serious form of HWD that must be promptly recognized as a surgical, rather than a medical, problem. When diagnosis of CS is delayed, the prognosis can quickly become grave.

This article focuses on the recognition and diagnosis of this life-threatening consequence of HWD. Visit tvpjournal.com to read Part 1 of this article series—Understanding Development of Caval Syndrome (November/December 2015).

DIAGNOSTIC OVERVIEW
The following scenario is all the history a veterinarian needs to place CS high on a diagnostic rule-out list:
1. It is springtime.
2. A dog presents to your hospital with acute weakness, and is breathing hard.
3. The dog was normal yesterday morning.
4. The dog is not receiving heartworm prevention.

These clues are often available to the clinician before entering the examination room. Although this diagnosis might be quickly ruled out during the physical examination, the fact remains: when the history is similar to the above description, CS should at least be a consideration.

Caval Syndrome: Disease Overview

Clinical signs of CS arise when adult heartworms move retrogradely from the pulmonary arteries to the heart. When this occurs, a mass of worms traverses the tricuspid valve and becomes wrongly located in the right ventricle, atrium, and often the vena cava. In this aberrant location, the worms can become knotted and tangled. As red blood cells regurgitate and squeeze through this tangled mass of worms, they are traumatized and lysed, leading to anemia, hemoglobinemia, and hemoglobinuria. Icterus may or may not be noted, but bilirubinemia secondary to hemolysis is typical.

Because the mass of worms traverses the tricuspid valve, the obstruction prevents valve closure and impedes blood flow through the right heart. Cardiac function is further hindered by obstruction of the distal pulmonary arteries by a high number of worms, as well as by endothelial proliferation, fibrosis, and embolic disease within capillaries, arterioles, and arteries. Impeded blood flow through these diseased vessels leads to increased pulmonary artery pressure, further compromising right-sided cardiac output.

Cardiac preload also increases, leading to congestion of the liver and spleen and, at times, the formation of ascites. Central lobular necrosis of the liver along with cholestasis has been described, and marked elevations of alkaline phosphatase, alanine aminotransferase, aspartate aminotransferase, and gamma-glutamyltransferase are common.

As one would expect, a reduction in right-heart output diminishes blood volume to the left heart, which, in turn, leads to left-sided output failure. Circulatory collapse ensues. Decreased peripheral arterial pressure results in reduced tissue perfusion. Metabolic acidosis and renal hypotension develop. Hypotension and hemoglobinemia potentiate compromised renal function, which is often represented by increases in blood urea nitrogen, creatinine, and phosphorus levels.

Active embolic disease and inflammatory changes within the pulmonary arteries and lung parenchyma may incite cough, but more commonly, respiratory signs of polypnea or dyspnea are related to circulatory failure, hypoxia, and respiratory compensation for severe metabolic acidosis. Inflammatory mediators may incite the consumption of coagulation factors and platelets, resulting in disseminated intravascular coagulation.

As one can see, CS is a complex disease involving both physical and physiologic components. Corruption of homeostasis occurs quickly, and the longer the heartworm mass inhibits cardiac function, the more likely death will ensue. Thus, surgical removal of this obstructive mass is the only means of reestablishing circulation and permitting homeostasis to normalize. Without surgical intervention, the short-term prognosis worsens; affected pets usually die within 1 or 2 days.

HISTORY

Pets presenting with CS have no or—at best—a poor history of receiving heartworm preventives. CS can occur throughout the year but is more common in the spring and early summer. This seasonality may be due to the timing and maturation of a high worm burden acquired during the previous summer.

Classically, clinical signs occur suddenly, when heartworms relocate to the right heart and cause obstruction. As such, the typical history is that of acute onset—clinically normal yesterday, sick today. Pet owners often report the following:

- Significant lethargy and weakness
- Anorexia
- Difficulty breathing
- Cough (occasionally)
- “Bloody” urine
- Pale gums
- Distended abdomen (less frequent).

CLINICAL FINDINGS

Physical examination usually reveals a lethargic, weak, and somewhat anxious pet experiencing mild to severe respiratory distress. Since the condition is usually acute at time of presentation, the patient may not yet be clinically dehydrated and often remains in good body condition. Other findings are listed in the Table.

DIAGNOSTICS

When CS is suspected, the diagnostic approach should be direct. The goal is to quickly confirm or refute the diagnosis by performing a urinalysis, heartworm test, and cardiac ultrasonography.

Urinalysis

When combined with history and physical findings, the urinalysis can single out CS as a top diagnostic consideration.

The most significant finding is a gross appearance of dark red to black coffee-colored urine, which is a result of severe hemoglobinuria—a consequence of intravascular red cell trauma and subsequent hemolysis. Although coffee-colored urine is considered by some to be pathognomonic for CS, any cause of rapid intravascular hemolysis could potentially
cause this clinical finding (Figure 2).

Other clinical findings include:

- Proteinuria, bilirubinuria, and hemoglobinuria (usually measured as hemolyzed blood)
- Sediment casts that indicate tubular disease and may occasionally reveal the presence of microfilaria.

<table>
<thead>
<tr>
<th>TABLE. Caval Syndrome: Clinical Findings</th>
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<tbody>
<tr>
<td>Easily visualized bounding jugular pulse</td>
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<td>Jugular distension</td>
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<tr>
<td>Lethargy and weakness</td>
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<tr>
<td>Mild icterus</td>
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<td>Mild to severe ascites</td>
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<tr>
<td>Mild to severe hepatomegaly and/or splenomegaly</td>
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<tr>
<td>Mild to severe respiratory distress</td>
</tr>
<tr>
<td>Normal or harsh lung sounds, with crackles sometimes noted</td>
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<tr>
<td>Pale mucous membranes with delayed capillary refill time</td>
</tr>
<tr>
<td>Pronounced tricuspid murmur</td>
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<td>Tachycardia</td>
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<td>Weak peripheral pulses</td>
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Heartworm Test

Pets with CS typically have a heavy heartworm burden; therefore, infection confirmation is not usually difficult.\(^2\),\(^3\),\(^8\) As previously noted, microfilaria may be found during evaluation of urine sediment, but more commonly, specific testing is required:

- Direct evaluation of a single drop of blood may reveal microfilaria
- Heartworm antigen test results are usually interpreted as “positive.”

If test results are negative, but history and clinical findings suggest the possibility of CS, additional steps must always be taken to definitively confirm or rule out this disease (see When Tests Say No but Other Findings Say Yes).

Cardiac Ultrasonography

CS is confirmed by ultrasonography when a mass of adult heartworms is identified within the right atrium, ventricle, and tricuspid orifice. Because the cuticular wall of the adult worm is reflective, it can be easily visualized as parallel, hyperchoic lines (Figure 3, page 66); a mass or tangle of worms is represented as numerous short, white, parallel lines (Figure 4, page 66) and, when observed, confirms the diagnosis.

I intentionally use the term cardiac ultrasonography rather than echocardiography because:

1. A complete assessment of the heart is not necessary; rather, the practitioner simply needs to identify the presence of adult heartworms in the right side of the heart to confirm CS.

When Tests Say No but Other Findings Say Yes

Interestingly, I have diagnosed pets with CS when both microfilaria and antigen tests have yielded “negative” results. This can be confounding when history and clinical findings make CS the most likely diagnosis. It also seems counterintuitive given the presence of a high population of mature adult worms typically composed of both sexes.

However, even in heavily infected animals, “negative” test results do not confirm the absence of heartworms. They simply indicate that:

- Microfilaria numbers are below detectable limits
- No adult heartworm antigen is detected.

Much has been written about possible reasons for “false-negative” results in heartworm-infected pets, but no specific explanation has been given for such findings in dogs with CS. Nonetheless, in my opinion, it makes sense that the role of the immune system and production of antibodies to various stages of heartworms can result in the arrested production or elimination of circulating microfilaria, and formation of antibody complexes with adult worm antigen, rendering them unavailable for detection by routine tests.

FIGURE 2. Hemoglobinuria depicted as a dark red to black, coffee-colored urine (A); urine can be collected via cystocentesis (B). This finding is often considered pathognomonic for CS. Courtesy Merial
The Role of Ultrasonography

Today, high-quality ultrasound equipment is found in many clinical practices, but just a decade ago this technology was far less available. Without ultrasonography, experienced veterinarians diagnosed CS on the basis of history, clinical findings, and minimal diagnostic tests. Confirmation was possible only when the mass of worms was surgically removed. Thus, a great deal of confidence was required to tell a pet owner that, while it could not be proven, there was a mass of heartworms blocking the pet’s heart and surgery was needed to blindly attempt to remove this mass.

Today, ultrasonography makes our job much easier, but we should keep in mind that many pets were diagnosed and underwent successful surgeries long before this technology was readily available. The inability to perform an ultrasound on a heart should not deter a clinician from diagnosing CS and performing surgery when findings strongly support the diagnosis.

2. For many practitioners, the thought of performing echocardiography can be intimidating; as a result, many of us fail to place an ultrasound probe on a heart.

Any practitioner with access to ultrasonography and minimal training can make this diagnosis (see The Role of Ultrasonography); it does not require the expertise of a cardiologist. As such, the practitioner can and should perform ultrasonography as soon as possible, keeping in mind that surgery would most appropriately be done that same day.

IN SUMMARY

Most veterinarians have great insight for diagnosing illness and, at times, can readily guess a diagnosis simply on the basis of history, such as in the following examples:

• An owner calls to report that her 6-week-old puppy is weak, with pale gums and black stool. Our usual thought is hookworm anemia.

• An intact female was in heat a month ago and is now anorexic, with a fetid vaginal discharge. We are quick to think of pyometra.

CS can be similarly suspected when history indicates an acute onset of respiratory distress and weakness in a pet that is not receiving heartworm prophylaxis. Add to the history a tricuspid murmur, bounding jugular pulses, and hemoglobinuria, and a clinician might well be on the way to diagnosing CS.

Because HWD is no longer limited to the southern and coastal regions of the U.S., it is important that we all become capable of recognizing its clinical signs. Being familiar with the history and presenting signs of CS allows clinicians the opportunity to save a life when a pet presents with this syndrome.

Read the next article in this series, Management & Treatment of Caval Syndrome—which will cover the medical management and surgical extraction of heartworms in canine CS—in an upcoming issue of Today’s Veterinary Practice.

CS = caval syndrome; HWD = heartworm disease

References


1. **History Findings**
- Presentation of patient in late winter or early spring
- History of incomplete heartworm prevention
- Acute onset of respiratory difficulty and weakness

2. **Physical Examination Findings**
- Polyphnea or dyspnea
- Pale mucous membranes (**Figure 5**)
- Jugular distension with bounding pulse (**Figure 6**)
- Distinct cardiac murmur suggestive of tricuspid regurgitation
- Palpable hepatomegaly and/or splenomegaly

3. **Diagnostic Findings**
- Dark red to coffee-colored urine (**Figure 2**)
- Microfilaremia and/or positive antigen test result (**Figure 7**)
- Cardiac ultrasonography confirming adult heartworms in the right heart (**Figures 3, 4, and 8**)

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**FIGURE 5**. Pet with CS; note pale mucous membranes and decreased capillary refill time. **Courtesy Merial**

**FIGURE 6**. The jugular vein is visibly distended in this patient, and a bounding jugular pulse is readily apparent. It may be necessary to shave the hair of a thick- or long-haired dog in order to observe this finding. **Courtesy Merial**

**FIGURE 7**. Pets with CS are often infected by a high number of heartworms; therefore, it is common to find many microfilariae even on direct evaluation of a single drop of blood. **Courtesy Merial**

**FIGURE 8**. Cardiac ultrasonography is the definitive test for CS. Adult worms are easily visualized within the right heart. **Courtesy Merial**

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