Cardiovascular disease, and especially aortic rupture, has recently gain attention in the sport horse community. Though aortic rupture and sudden cardiac death can occur sporadically and without warning, recognition of clinical signs associated with aortic rupture and pre-existing aortic aneurysm and knowledge of appropriate treatment recommendations and prognosis for these horses are critical for the sport horse practitioner.

Important points:

- Aortic rupture can occur with or without pre-existing aortic aneurysm
- Sinus of Valsalva aneurysms could be linked to congenital connective tissue disorders such as DSLD or HERDA
- Aortic rupture occurs most frequently at the aortic root (Sinus of Valsalva) resulting in an aorto-cardiac fistula (Friesians are the exception, and tend to experience aortic rupture at the ligamentum arteriosum, resulting in a aorto-pulmonary fistula)
- Clinical signs of aortic root rupture include a bilateral continuous machinery murmur, signs of acute distress and pain (generally mistaken for colic), and sudden death
- Intact aortic root aneurysms are typically clinically silent, though they may have a murmur of right outflow tract obstruction or aortic regurgitation
- Treatment of aortic root rupture includes management of acute congestive heart failure (inotrope, ace-inhibitor, diuretic) and arrhythmias
- Treatment of intact aortic aneurysm includes management of its sequelae, including aortic regurgitation
- If a horse survives the initial aortic rupture incident, prognosis for life is variable and dependent on the size and location of the rupture, however, these horses will eventually die of congestive heart failure
- It is COMPLETELY contraindicated to ride, drive, or otherwise work horses with either aortic rupture or an intact aortic root aneurysm, due to increased risk of ventricular arrhythmias, collapse, and sudden death

Aortic anatomy:

The anatomy of the aorta and aortic root is important in understanding both the clinical signs associated with aneurysm and rupture, and their consequences. The aortic valve is comprised of three semi-lunar cusps; a non-coronary cusp, a right coronary cusp, and a left coronary cusp. The sinutubular junction is the junction between the ascending aorta and the semilunar cusps comprising the aortic valve. The annulus fibrosis is the junction between the aortic valve and the left ventricle, and the dilation between the sinutubular junction and the annulus fibrosis is the Sinus of Valsalva (SOV). There are three of sinuses associated with each of the cusps, a right coronary sinus, left coronary sinus, and a non-coronary sinus.
The ascending aorta is connected further proximally to the pulmonary artery by the ligamentum arteriosum.

![Diagram of the aortic root](image)

Figure 1. Anatomy of the aortic root (Charitos et al, 2013)

**Aortic rupture:**

Aortic rupture in most horses occurs at the aortic root (Sinus of Valsalva), resulting in an aorto-cardiac fistula. However, aortic rupture in Friesians occurs in a different location than in other breeds, near the ligamentum arteriosum where the pulmonary trunk and wall of the aorta connects. This results in an aorto-pulmonary fistula, instead of an aorto-cardiac fistula.

Aortic root rupture was first described in breeding stallions in 1967 by Rooney et al. The majority of deaths in this study occurred during or shortly after breeding, and therefore aortic rupture was thought to be associated with a combination of elevated blood pressure and recoil force on an anatomic weak point in the aortic root. However, more recent publications have described breeding stallions which died at rest, and not during strenuous exercise such as breeding, suggesting that elevated blood pressure during breeding is not the only cause of rupture.

Rupture of the aortic root is thought to be a rare condition, comprising only 3/984 deaths in young race horses according to one study, though the prevalence in other sport horses is unknown, and this study was performed in young racehorses, which is not a common population for aortic rupture.

**Aortic aneurysm:**

Aortic root rupture can occur with or without the presence of an aneurysm, and case reports have described horses both with evidence of aneurysmal dilation prior to rupture, and dissection through the aortic ring without aneurysm.

An aneurysm is a dilation which can have both congenital and acquired causes. Aneurysms of the aortic sinus in both humans and horses occur most commonly in the right Sinus of Valsalva. These aneurysms can lead to rupture and dissection into the interventricular septum, or formation of an aorto-cardiac fistula (connection between the aorta and the heart chamber).
The prevalence of intact aneurysm in the general population of horses is unknown, especially as most intact aneurysms are clinically silent. Like humans, SOVA’s are most common in males, occur most commonly in the right coronary sinus, and most commonly rupture into the right ventricle.

**Pathogenesis of SOV aneurysm:**

It is likely that SOV aneurysms in horses are both congenital and acquired. Congenital SOV aneurysms are caused by a complete lack of fusion of the aortic media and annulus fibrosis, with subsequent dilation. Acquired aortic aneurysms, which occur due to weakness of the smooth muscle (medial layer) of the aorta with progressive dilation, have also been reported in the literature.

Potential mechanisms for weakening of the aortic media and acquired aortic aneurysm include connective tissue disorders, infection, and degenerative disease. One connective tissue disorder recognized in horses with potential aortic involvement is degenerative suspensory ligament desmitis (DSLD). DSLD is a disease that occurs mostly in Peruvian Paso’s, and involves abnormal deposit of proteoglycans, and abnormal collagen within ligaments. In a study by Halper et al in 2006, histologic examination of other body systems in these horses revealed evidence of these proteoglycan deposits and abnormal collagen fibrils within the media of the aorta support that a connective tissue disorder could lead to weakening of the aortic media and aneurysm formation. Studies have not been performed investigating the occurrence of aneurysm or aortic rupture in Peruvian Paso’s or in other horses with DSLD in comparison with other horses.

Hereditary Regional Dermal Asthenia (HERDA) in Quarter horses is a disease which results in abnormal, thin, fragmented collagen within the deep dermis, and is similar to Ehlers Danlos syndrome in people. This type of collagen disorder could also lead to aortic aneurysm, as does Ehlers Danlos in people, however specific histologic studies of the aorta in these horses have not been performed.

**Clinical signs:**

Horses with aortic root rupture and secondary aorto-cardiac fistulas typically present with classic signs of acute distress and pain, which is commonly mistaken for signs of colic. Acute distress and pain can be related to the rupture itself, pain associated with dissection of blood through the interventricular septum, or secondary to the presence of ventricular tachycardia. These horses have a classic bilateral continuous machinery murmur which is loudest on the right, will have bounding pulses due to rapid diastolic runoff, and often have ventricular ectopy. Sudden death in these horses has been linked to ventricular arrhythmias that occur as blood dissects through the interventricular septum and disrupts the conduction tissues.

Intact SOV aneurysms are thought to be primarily asymptomatic. These asymptomatic aneurysms are generally discovered when a murmur of right outflow tract obstruction (systolic murmur with point of maximal intensity over the pulmonic valve) or aortic regurgitation (diastolic murmur with point of maximal intensity over the aortic valve) is ausculted. Aortic regurgitation in these cases can occur due to lack of support of the aortic annulus secondary to aneurysm formation. Though horses with intact aneurysms are generally thought to be asymptomatic, horses can eventually show clinical signs of congestive heart failure secondary to significant aortic regurgitation.
**Diagnosis:**

Diagnosis of aortic root rupture in the horse is made by echocardiographic examination after clinical suspicion.

![Image](image1.png)

Figure 2. Horse with intact right Sinus of Valsalva aneurysm bulging into the right outflow tract

![Image](image2.png)

Figure 3. Horse with rupture of the aortic root and development of aorto-cardiac fistula with communication of the aortic root and right ventricle

**Treatment:**

Unlike humans, the mainstay of treatment of aortic root rupture in horses is medical management of its sequelae, namely congestive heart failure. Like all animals, the primary tenants of treatment of congestive heart failure include administration of an ace inhibitor, diuretic, and positive inotrope.
Surgical treatment of SOV aneurysms and aorto-cardiac fistulas in horses like those done in humans have obvious limitations, including visibility, lack of cardiopulmonary bypass machines large enough for horses, and graft size availability. Successful trans-catheter closure of an aorto-cardiac fistula with an Amplatzer septal occluder has been described in one previous case report by Javiscus et al in 2010, though this horse was euthanized 45 days after presentation due to clinical deterioration.

**Prognosis:**

The prognosis for horses with intact SOV aneurysms is difficult to determine, as the majority of intact aneurysms are clinically silent. However, the outcome of these cases is likely dependent on the presence of secondary cardiac disease such as aortic regurgitation. There is also a lot of variability in the prognosis for horses with ruptured aortic aneurysm. In a study by Marr et al in 1998, horses were reported to survive anywhere between a day and 12 months or more, and horses both here at Penn and at Davis have survived up to 4 and 6 years with a ruptured aneurysm respectively. It is likely that if a horse survives the initial rupture event, they will survive weeks to months, but most commonly these horses would develop congestive heart failure from biventricular volume overload.

Prognosis for life in both horses with both aorto-cardiac fistulas and intact aortic aneurysms is poor, and return to work is completely contraindicated. This is due to the increased risk of development of ventricular arrhythmias, collapse, and sudden death, making them a risk to themselves, their handlers, and their riders/drivers.

**References:**


Halper, J, Byoungjae, K, Khan, A., Hae Yoon, J. and Mueller, E “Degenerative suspensory ligament desmitis as a systemic disorder characterized by proteoglycan accumulation” BMC Veterinary Research (2006)


