The “Big Horse” Disease

*Draft and warmblood breeds are prone to the muscle disorder EPSM. Here’s what you need to know.*

By Eleanor M. Kellon, VMD

**Equine polysaccharide storage myopathy, or EPSM (also known as PSSM or EPSSM), is a muscle disorder of glycogen metabolism that was first described in the 1990s. It quickly became a bit of a “disease du jour,” taking the blame for everything from all cases of tying-up (equine exertional rhabdomyolysis) to gait abnormalities.**

Although EPSM remains something of a scapegoat, it is a real potential issue for many horses. Dressage enthusiasts need to familiarize themselves with this disease because our sport’s most common breeds, the warmbloods, are in the high-risk group for EPSM.

In this article, I’ll describe the causes and symptoms of EPSM; then I’ll explain how the disorder is diagnosed and how it’s being treated successfully with diet and exercise.

**Sugar, but Not Sweet**

The “P” in EPSM stands for polysaccharide, which is essentially a chain of sugars. Hay, other forage sources, and the starch in grain feeds contain glucose, a simple sugar or monosaccharide. The horse’s muscles and other tissues store glucose in the form of glycogen. Glycogen fuels the body’s activity during exercise; performance suffers when glycogen stores are depleted.

In horses with EPSM, something goes wrong with the body’s glycogen-storage system. There are two types of EPSM.

**Type 1 EPSM** is characterized by accumulations in the muscles of an abnormal polysaccharide that differs from normal glycogen. This abnormal polysaccharide cannot be broken down by the body’s enzymes to provide the muscle cells with needed energy.

Horses with type 1 EPSM have a genetic mutation in an enzyme called glycogen synthase 1, or GYS1. Glycogen has a “backbone” of a straight train of glucose molecules, with branches off that straight chain like the branches of a tree. In type 1 EPSM, the glycogen backbone is believed to lengthen so quickly that the formation of side branches cannot keep up. The resulting abnormal form of glycogen is more resistant to being broken down because the muscles’ enzymes normally act on the ends of the branches. Because the muscles are not getting enough energy, they can neither perform normally nor relax normally after contracting.
Type 2 EPSM occurs when the muscle tissue stores excessive amounts of glycogen in abnormal locations. The reason for this is currently unknown.

In general, horses with EPSM do not have trouble breaking down the normal form of glycogen. However, they do so very inefficiently. As a result, horses with EPSM produce more lactic acid, have decreased numbers of mitochondria (the “powerhouses” of the cells for generating energy aerobically), and are less able to recycle the metabolites of ATP (adenosine triphosphate, the energy currency of cells) back to ATP.

Regardless of the type, the bottom line with EPSM is that affected horses do not use glucose, whether blood glucose or glucose stored as glycogen, efficiently. The result is that the muscles are either too weak to contract at all or are unable to relax if they do contract (relaxation of muscle also requires energy).

Symptoms

Symptoms of EPSM vary tremendously. Some horses—typically the draft breeds—may have attacks so severe that they cannot rise, and weakness is the predominant symptom.

Aerobic vs. Anaerobic

Muscles can generate energy from glucose (glycogen), fats, and protein. To burn fats and protein, the cells’ mitochondria (their “power plants”) need oxygen; this is known as aerobic energy generation.

The mitochondria can also burn blood glucose (or glucose from stored glycogen) anaerobically—without oxygen. This is a rapid way to generate energy, but it is very inefficient compared to aerobic energy generation.

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Many horses with EPSM experience tying-up (exertional rhabdomyolysis), which is an exercise-induced breakdown of skeletal muscle. The horse begins by showing a reluctance to go forward, eventually coming to a complete stop. The muscles are hard and painful to the touch, particularly in the hindquarters. The horse is in obvious distress, sweating and blowing and sometimes pawing. In severe cases, the muscle pigment myoglobin stains the urine, turning it red to brown in color.

A variety of gait disturbances have been reported in EPSM horses. These include stiffness, choppy or jerking movements, tripping, and difficulty engaging the hindquarters. The horse may have difficulty holding up the hind legs for the farrier, often with trembling of the limbs.

Muscle fasciculations (muscle twitching) may be present at rest in

Shivers and EPSM

Shivers is a neurological and muscular disorder of one or both hind legs. It may begin as exaggerated snatchng movements when a person attempts to handle an affected horse’s back feet, and it progresses to episodes in which the leg is held in a flexed position with the muscles visibly quivering. The tail also elevates and quivers to varying degrees. Backing reliably triggers signs, as may trying to work on the hind feet.

Because shivers is most common in some of the breeds that are at higher risk for EPSM, such as drafts and warmbloods, it has been suggested that shivers is caused by EPSM. However, a study of Belgian draft horses by researchers at the University of Minnesota found no direct link between the two conditions. A horse can have both shivers and EPSM, but the correlation appears coincidental.
a horse with EPSM. Many affected horses experience muscle loss, difficulty developing good muscling, or both, especially along the topline and rump. Possible conformational changes include the hindquarters becoming more sharply sloped and the pelvis’s appearing “tucked under.”

Although full-blown tying-up episodes may occur in the warmblood breeds, these horses are likely to show more subtle signs, such as muscle tightness, sore backs, difficulty engaging, and difficulty progressing in training. Owners of horses diagnosed with EPSM may note a poor tolerance for work or an apparent reluctance to work.

**Diagnosing EPSM**

EPSM cannot be diagnosed by symptoms alone, even if the horse ties up.

A blood-chemistry panel may show elevated muscle enzymes, with mineral abnormalities if the horse tied up recently. However, these findings do not constitute a positive diagnosis for EPSM.

A genetic test using blood or hair can diagnose type 1 EPSM. Muscle biopsy with special staining can diagnose both type 1 and type 2 EPSM.

**Treatment**

Maintaining a regular exercise schedule with as much turnout as possible is an important part of the management regimen for EPSM, as it helps to prevent stiffness. More important, exercise stimulates the burning of carbohydrates for fuel rather than storage as glycogen.

A low-carbohydrate diet with supplemental vitamin E and selenium and added fat is the standard treatment for EPSM. This dietary regimen works well for many horses but can take several months to take full effect, during which time they may relapse.

There are different approaches to calculating the appropriate amount of fat to add to the horse’s diet. A standard rule of thumb is to feed 1 lb. of fat per 1,000 lbs. body weight. Research-
ers at the University of Minnesota recommend determining how many calories the horse needs to maintain his optimum body weight. They suggest feeding hay that is low in sugars and starches, a ration balancer, and fat. Their studies of Quarter Horses with EPSM led them to recommend feeding fat as 6 to 10 percent of the total ration by weight (usually 1 to 5 lbs. of rice bran or up to 600 ml of oil per day). A low-starch, high-fat concentrate may be used if the horse’s workload is heavy enough to justify feeding the extra calories.

For the last five years I have been working closely with owners of EPSM horses to develop an alternative to the high-fat diet. Some horses develop insulin resistance and even laminitis as a result of a high-fat diet. Others refuse to eat the large amount of fat or show only a partial response to the high-fat diet.

The core of the alternative EPSM diet is the same as for high-fat feeding: low sugar and starch from tested hay, and “bucket feeds” that are low in sugar and starch, such as beet pulp. Minerals are tested, and deficiencies or imbalances are corrected. Adequate salt intake is guaranteed by adding it to the feeds, as even slight dehydration has a negative effect on muscle function. Vitamin E completes the base diet.

Instead of feeding fat, the alternative EPSM diet includes supplemental acetyl-L-carnitine at 1 gram per 100 lbs. of body weight. L-carnitine is a substance that is naturally produced by the body. It is essential for carrying fats into the mitochondria to be burned as fuel. Acetyl-L-carnitine is also an antioxidant, stimulates the production of mitochondria by cells, and directs glucose away from glycogen synthesis and into energy pathways.

With this approach, improvement may be seen as rapidly as three to four days after starting a horse on acetyl-L-carnitine. The horse’s movement and ability to tolerate work improve over the next few weeks.

Test and Control

EPSM can severely interfere with a dressage horse’s exercise tolerance and ability to progress in training. If you suspect that your horse has EPSM, do the diagnostic testing, as this disorder cannot be diagnosed by symptoms alone. Fortunately, EPSM can be controlled effectively. Regular exercise is a key component in management. Feeding a well-balanced diet that is low in sugar and starch with supplemental fat or acetyl-L-carnitine is therapeutic.

Eleanor M. Kellon, VMD, operates Equine Nutritional Solutions in Robesonia, PA. Her courses on equine nutrition and other topics (listed at DrKellon.com) are USDF University-accredited.

Dr. Kellon is the author of several books, including The Older Horse and Guide to First Aid for Horses. A former contributing editor to Horse Journal, she is now a staff veterinarian at Uckele Health and Nutrition Inc.