

## Polysaccharide Storage Myopathy—Unraveling The Mystery

*A high-fat, low carbohydrate diet turns some horses who suffer from this syndrome from miserable to magnificent.*

Erin Hartly

Two years ago, Polly Merrill thought she was at the end of the road with Telemakhos, or “Mikey,” an 8-year-old Thoroughbred she’d hoped to event.

“I was either going to turn him out or put him in the ground,” said Merrill, of Sherborn, Mass.

For the two years she’d had him, every day had been a struggle. He was so unruly that she wore a helmet and gloves every time she handled him. (And he still managed to break six of her ribs and give her a concussion.) He couldn’t stand to be confined—he’d kick at the walls of his stall and refused to stand in the crossties.

He had to be sedated for the farrier. He’d stand still in one spot in his turnout paddock and buck. He couldn’t canter. He couldn’t jump a combination of crossrails without crashing through one of them. He couldn’t maintain a frame while walking uphill. He spent about 20 minutes of every hour lying flat on his side in the pasture. He was skinny and hard-eyed and, in a word, miserable.

Still, Merrill adored the horse and felt sure that there was a physical cause for his issues. She and her veterinarian ran through the usual diagnostics—lameness exams, bloodwork, scoping for ulcers, testing for Lyme disease. They tried massage and acupuncture. Nothing helped.

“It was going on and on and on, and I was getting more and more broke,” Merrill said. Her veterinarian finally threw up her hands, told Merrill that she’d exhausted all of her theories, and suggested she get on the Internet and see what she could find.

After describing Mikey’s symptoms on *The Chronicle of the Horse’s* Internet bulletin board, Merrill was referred to a

**“My greatest reward is to have people who knew Mikey then see him now. They can’t believe he’s the same horse,” said Polly Merrill after treating him for equine polysaccharide myopathy.**

website about equine polysaccharide myopathy, a syndrome primarily characterized by increased storage of glycogen (a form of stored glucose for energy) in muscle cells. The symptoms—vague hind end lameness/weakness, trouble performing, abnormal gait—sounded similar enough to Mikey’s symptoms that Merrill brought up the possibility with her veterinarian.

“She looked at me and said, ‘That’s only seen in draft horses,’” Merrill said.

But Merrill decided to go ahead and switch Mikey to the high-fat, low-starch diet recommended for horses with polysaccharide storage myopathy. Couldn’t hurt, might help, she figured. And there was little else left to try.

Two years later, Merrill rode Mikey in his first three-day event, at the Morven Park CCI\* (Va.). Although they retired on cross-country after a green mistake and a fall early in the

course, Merrill was buoyant—considering the horse Mikey used to be, it was a stunning victory that they’d been able to compete at all.

“I finished phase B and asked my daughter, ‘Do I have bugs in my teeth?’ because I was grinning so much,” she recalled with a laugh.

“My greatest reward is to have people who knew Mikey then see him now. They can’t believe he’s the same horse,” she said.

### ► Not A Simple Solution

Merrill is one of a growing number of sport horse owners who believe polysaccharide storage myopathy is to blame for their horses being not-quite-right, and who have found that diet changes bring about remarkable results.

But the disorder is surrounded by considerable confusion. Increased glycogen storage has been documented in a wide variety of breeds, but certain breeds seem to show different symptoms than others.

It’s not yet confirmed that the exact same disease is responsible for the various symptoms that

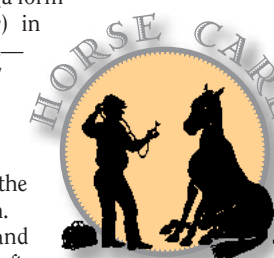
seem to be shown across different breeds, although experts believe it’s likely.

There’s also some disagreement over the diagnosis. Polysaccharide storage myopathy can be diagnosed through a muscle biopsy (see sidebar), but many owners, like Merrill, opt to change a horse’s diet without doing the biopsy—if the horse gets significantly better, it’s often presumed to have the disease, although there are other muscle disorders that also seem to respond to high-fat diets.

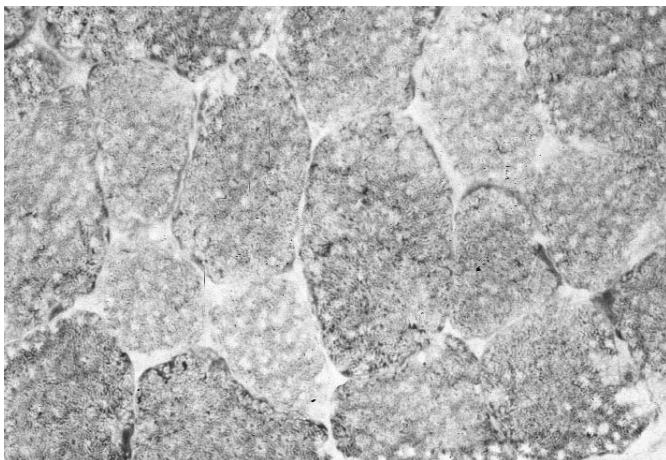
And if an owner does elect for a biopsy, opinions differ about what the biopsy must show to conclusively diagnose polysaccharide storage myopathy. Of the two primary U.S. facilities for diagnosis, one looks for increased glycogen storage, and the other looks for increased glycogen storage in addition to an abnormal polysaccharide. Both look for abnormal glycogen storage and storage of an abnormal indigestible polysaccharide. But one will make the diagnosis based on abnormal glycogen alone, and the other will only diagnose EPSM/PSSM if there is also an abnormal indigestible polysaccharide.

Even the name of the disorder can vary. Some refer to it as equine polysaccharide storage myopathy, abbreviated EPSM or EPSSM, and some call it simply polysaccharide storage myopathy, or PSSM.

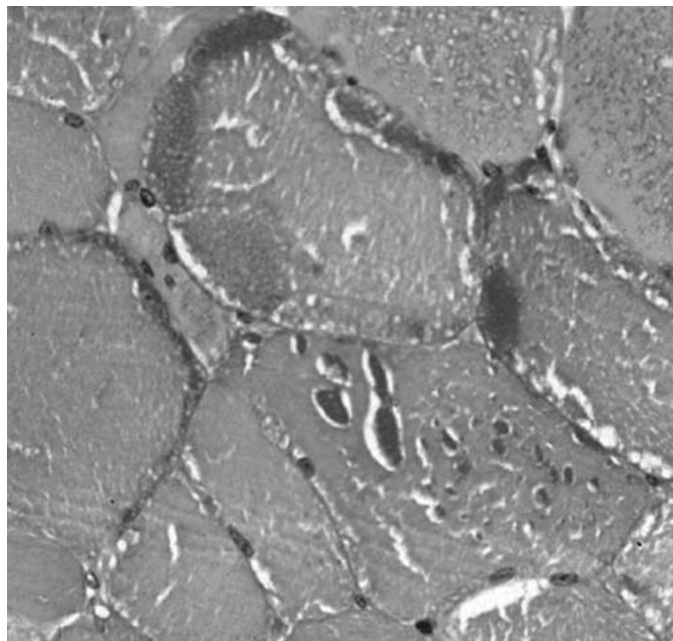
EPSM/PSSM only entered the medical consciousness in the late 1980s when Dr. Stephanie Valberg, currently director of the University of Minnesota Equine Center, first noted an anomaly in the storage of glycogen in the muscles of Quarter Horses that suffered



(Gary Cappage/CRC Photography Photo)



**Above:**The normal glycogen pattern is fine granules distributed relatively evenly throughout the fibers.



**Right:** While the abnormal pattern forms large aggregates.

from tying-up. She called the disease PSSM.

A few years later, Dr. Beth Valentine of Oregon State University recognized the same excess glycogen storage in a group of draft horses that also suffered from tying-up, and dubbed the disorder EPSM. Both veterinarians have continued to study the disease in detail and are considered the foremost experts on the disorder in the United States.

Valberg has found that Quarter Horses with EPSM/PSSM show typical tying-up symptoms—hard muscles, pain and stiffness, sweating and reluctance to move, especially when exercised after a period out of work. But she's also found that warmbloods with EPSM/PSSM exhibit tying-up symptoms less frequently and instead seem to suffer from gait abnormalities, lack of impulsion and muscle soreness.

Her observations on warmbloods are sim-

ilar to Valentine's findings in draft horses with EPSM/PSSM—occasional tying-up, but affected horses more frequently show other symptoms, like loss of muscling, hind end lameness, lack of energy, poor performance and abnormal gait.

Both researchers have found that horses with EPSM/PSSM respond well to a diet that limits starches (primarily by eliminating grain) and provides additional fat, as well as regular turnout and exercise.

### ▶ A Puzzling Syndrome

Researchers are slowly sorting out the pieces of the EPSM/PSSM puzzle, but they aren't yet certain exactly what causes the disorder or why affected horses exhibit the symptoms they do. What is clear is that horses with

EPSM/PSSM are metabolically different than normal horses, Valentine explained.

"Something is wrong with the way [EPSM/PSSM horses] are generating energy in the muscle—they're not generating enough, they're not generating it fast enough, and they hit an energy wall," Valentine said.

A horse's natural diet—grass—is low in starch and high in fiber. As athletic demands on horses increase, grain is often added to the diet as an additional source of starch, and therefore, energy. When a horse consumes a grain meal, the carbohydrates in grain are broken down and absorbed through the lining of the small intestine as simple sugars, mainly glucose. Cells can pull glucose from the blood and convert it into energy.

In order to keep a relatively constant level of blood glucose available, horses (and humans) rely on hormones to pull glucose from the blood when it's plentiful and release it back into the bloodstream when levels fall too low.

Insulin is the hormone that triggers the removal of excess glucose, which is converted and stored in muscle and liver cells as glycogen. When the levels of glucose in the blood fall again, another hormone signals for glycogen to be broken back down into glucose.

Valberg has found that horses with EPSM/PSSM are more sensitive to insulin than normal horses—in essence, their metabolisms are more effective at converting glucose to glycogen, leading to more storage of glycogen in muscle cells than would be seen in horses that don't suffer from EPSM/PSSM.

While EPSM/PSSM horses have trouble metabolizing starch, they have no such issues converting dietary fat to energy. So when these horses are given a diet lower in starch and higher in fat, their muscle cells are prompted to use fat as an energy source instead of starch, and usually, the symptoms associated with EPSM/PSSM lessen or disappear completely.

Precise diet recommendations differ, depending on whom you ask, but the main tenet is that a horse suffering from EPSM/PSSM should not be fed traditional grains or sweet feeds because of the high starch content.

Valentine recommends that a diet for an EPSM/PSSM horse provide 20 percent of the daily calories from fat, and an equivalent of 2 cups of oil (1 lb. of fat) per 1,000 lbs. body weight per day. She recommends a low-starch feed like alfalfa pellets or beet pulp in place of traditional grain.

Valberg, on the other hand, recommends tailoring the diet to the severity of a horse's symptoms and its body condition. "The amount of starch they can tolerate will vary," she said, adding that she's found a wide variety among Quarter Horses she's studied. "The more severe the signs, the lower the amount of starch they can tolerate."

She doesn't recommend a minimum level of fat supplementation but suggests feeding rice bran (which is high in fat), oil, or a specialized low-starch pelleted feed like Re-leve, which she developed for Hallway Feeds. A diet that supplies about 13 percent of daily calories from fat has been sufficient to abolish signs of EPSM/PSSM in the worst cases Valberg has studied.

Both experts say high quality hay and forage are also an integral aspect of any EPSM/PSSM horse's diet.

Although the diet change tends to get most of the attention, Valberg emphasizes the importance of the complete, recommended approach: regular exercise, taking care to increase the level of work slowly, proper conditioning, regular turnout, weight control, and a low-starch/high-fat diet, including proper vitamin/mineral balance.

### ▶ Isn't That Something Draft Horses Get?

Both Valberg and Valentine have documented the disorder in a range of breeds, but there's still a common perception that EPSM/PSSM is a "draft horse thing," which perhaps explains why knowledge of the disorder is sometimes lacking in the sport horse world. Often, it's not until every traditional solution for not-quite-right horses has failed—as in Merrill's case—that owners and veterinarians get desperate, start grasping at what they think are straws, and stumble across EPSM/PSSM.

"We see more and more muscle samples coming from sport-horse-exclusive [veterinary] practices," Valentine said. Veterinarians who are initially skeptics are often astonished at the dramatic improvement made by the majority of EPSM/PSSM horses when diet changes are implemented, she added. "Once they've seen that, they start recognizing [EPSM/PSSM] in horses they've seen for years."

The situation Valentine describes is exactly what Merrill experienced. Although her veterinarian initially greeted her suggestion of EPSM/PSSM with skepticism, Mikey is now a "poster boy" for EPSM/PSSM at the Massachusetts Equine Clinic because

of his dramatic turnaround, Merrill joked.

"A lot of people are struggling with these horses for years and years. For most of them, it's a relief to have an answer," Valentine said.

Although EPSM/PSSM cannot be cured—affected horses will always show abnormalities in a muscle biopsy—the good news for owners is that the disorder can be managed into near non-existence with proper diet and exercise. Valberg has found that more than 95 percent of horses stop tying up with treatment. Valentine has likewise noted a response rate of about 90 percent, although she has observed very serious cases in draft horses that did not respond to treatment.

"Most horses respond remarkably to diet change," Valentine said. "They're so responsive, as a matter of fact, it's sort of mind-boggling."

Merrill's horse has been on a diet of alfalfa pellets and 3 cups of oil per day for two years, and he's a completely different horse, in both personality and performance. "Every one of those symptoms is gone," she said.

Mikey has been competing at preliminary level successfully, and he's become enough of a puppy dog on the ground that even Merrill's non-horsey husband is comfortable working with him.

The diet change isn't without its own challenges, however, especially for owners who try to implement Valentine's recommended high-fat regime. Some horses simply refuse to eat oil, or won't eat the recommended quantities.

When Merrill first presented Mikey with his new ration, you'd have thought she offered up a bucket full of poison, she said. With persistence and unwavering resolve, Merrill convinced him it was not only edible but was the only meal he was going to be offered, and now he licks his bucket clean without complaint.

Owners who are just trying to find higher fat/lower starch feed alternatives also have their work cut out for them. Feed labels, the content of which is regulated by federal law, don't include information on starch content. So carb-conscious horse owners have to call feed companies individually and ask. The good news is that feed companies are well aware of the demand for higher fat and lower starch feeds and have information and suggestions at the ready.

"I'm so pleased because I know for a fact that you could contact feed companies 10 or 15 years ago and ask 'What's the starch or sugar content?' and they'd say 'Huh?'" Valentine said. 🐾

## A Special Challenge for Sport Horses

Although most horses with polysaccharide storage myopathy seem to respond well to diet change, highly successful sport horses aren't "most" horses. Can equine athletes reach the upper pinnacles of performance with this particular handicap?

"A few might not be top competition horses," said Dr. Beth Valentine. "They might not be able to handle the stress. But for most horses, it has not proven to be a major stumbling block. Sometimes, they may be better [than before]."

For eventer Polly Merrill, her horse's condition means that she pays special attention to building up his hind end. Before Telemakhos, nicknamed Mikey, was diagnosed, he had some significant muscle wasting in his right hindquarter, so it took time to build up his strength, and Merrill makes sure to keep his hind end built up with careful hill work. Still, she said he's not as muscled as she'd like him to be.

Mikey also displays a gait abnormality typical of some EPSM/PSSM horses—a slapping motion with his right hind—and consistently moves shorter with that leg. He was actually held in the first jog at the Virginia CCI, and Merrill had to explain his history and show the ground jury how he's muscled differently on his right side than his left.

"Every dressage test says he needs to be more through," she admitted. "He will never be a beautiful mover."

But despite Mikey's early difficulties over fences, jumping poses no problem anymore. "[He] can jump anything," Merrill enthused. "It makes me cry."

Of particular interest in the sport horse world is the possibility that EPSM/PSSM is an inherited trait. Assuming horses with EPSM/PSSM are still able to achieve competitive success, is it wise to breed them?

According to Dr. Stephanie Valberg, no. She believes that EPSM/PSSM is almost certainly a genetically linked trait in Quarter Horses, and possibly in drafts and warmbloods as well. She's currently involved with research to develop a genetic test for EPSM/PSSM in Quarter Horses, which she hopes will enable Quarter Horse breeders to easily identify horses that carry the trait by simply pulling a few mane hairs and sending them off for testing.

Valentine holds an opposing view. "My take is that we have been breeding them. We've been breeding them for 100 years. We may have selected for this type of horse, and when their muscles are working well, they can be very high-performance horses," she said.

The disorder is so common in draft horses—Valentine believes up to two-thirds of drafts have it—that eliminating those horses from the breeding pool wouldn't leave many other candidates.

"Providing a horse responds to diet change and has other good attributes to pass on, I don't try to discourage [breeding]," she said.