

MSUD Explained

Understanding the basics of Maple Syrup Urine Disease.



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I hope everyone is getting some relief in terms of weather. The winter we have endured surely has been trying, and I would like to thank all of our Hereford breeders for their resiliency and dedication.

Recently, a genetic defect known as Maple Syrup Urine Disease (MSUD) was identified in the breed. I would like to take the time to answer some recurring questions concerning this disease.

What is MSUD?

MSUD is a lethal genetic abnormality and is a simple autosomal recessive gene. Affected calves have a defect in an enzyme that breaks down complex amino acids in the diet. The resulting buildup of these amino acids in the body causes lethal brain damage.

What are the symptoms of MSUD?

Affected calves are typically born without symptoms, but by 2 to 4 days old become slow, dull and eventually recumbent. The calf will often throw its head back and lie on its side, unable to rise. These calves may have some swelling of the brain at autopsy, but diagnosis requires laboratory investigation. This disease is also found in humans and is named for the smell of urine observed in human babies — the smell is not always noted in calves.

What is the frequency level of identified carriers for MSUD?

At this time, less than 1 percent of the 33,000 animals genotyped with the marker test for MSUD have been identified as carriers.

Will an animal identified as a carrier always sire a carrier progeny?

No, a carrier animal has a 50 percent chance of siring a carrier animal and a 50 percent chance of siring a noncarrier.

Can a calf have MSUD if only one parent is a carrier for MSUD?

No, for a calf to have MSUD, both parents must be carriers. Even then, there is only a 25 percent chance of having an affected calf. Consequently, this mating profile would also yield a 25 percent chance of an MSUD-free animal and a 50 percent chance of an MSUD carrier.

Did the American Hereford Association (AHA) pick and choose which animals were initially tested for MSUD?

No, the AHA published all results found for MSUD.

Why do some of my genotyped animals have a MSUD result while others do not?

The marker test for MSUD was only available on AHA genotype panels from roughly the fall of 2014 through the fall of 2017. Animals previously genotyped outside this timeframe are eligible for a standalone MSUD test at \$18. These requests must be made through AHA Customer Service.

Going forward, how will MSUD testing be handled when an animal is DNA tested?

The AHA has moved to a higher density genotype panel that will return MSUD results along with other abnormality results and a genomic profile. This genotype panel will allow for continued genetic advancement of the breed.

How can I identify animals in my herd that warrant testing for MSUD?

Members can run a potential defect carrier job for MSUD or any of the genetic abnormalities through MyHerd. This job can be accessed under the “Jobs & Reports” tab. For more information on how to run this job and

to receive a report for MSUD, please use this link:

Iorad.com/player/1553175/How-To-Run-The-Potential-Defects-Carriers-Report#trysteps-1

When I ran the MSUD carrier report, no animals came up as potential carriers. Does this mean I do not have any animals that could be carriers?

Not necessarily, because not all genotyped animals have a result

for MSUD. With continued testing for MSUD, there will likely be additional animals identified as carriers, so it is best to run the defect carriers report periodically.

Where can I access a list of animals identified as MSUD carriers?

Members can access a list of MSUD carrier animals at Hereford.org under the “Genetic Abnormality” tab under “Genetics.”

Reports of MSUD to the AHA and to the diagnostic lab at the University of Nebraska-Lincoln are absent in recent years. With an estimated gene frequency of less than 1 percent, the current occurrence of calf losses is estimated at less than 20 per 1 million births and is likely going unrecognized. Even with this low incidence rate, testing of at risk animals should be completed to ensure this abnormality does not propagate. **HW**

Genomics Continue to Advance Our Understanding of Calf Diseases

by **David Steffen**, DVM, Ph.D., ACV, University of Nebraska-Lincoln

Modest cost sequencing, the ability to cheaply and rapidly genotype tens of thousands of markers, and large genomic databases available to researchers have advanced industry approaches to investigating potentially heritable diseases. Decades ago, investigation was long, costly and often uncertain — breeders may have been reluctant to report problems, and breed groups considered risk and cost when confronted with abnormal calves.

Current technology allows modest investments to enlighten producers' understanding of diseases and can rapidly place tools in the hands of breeders to mitigate impacts and losses associated with deleterious genotypes. Cost benefit analysis is still prudent in prioritizing projects for funding, but benefits and cost reductions allow for affordable investigation of more cases.

Investigation

Current investigations into several diseases at the University of Nebraska-Lincoln (UNL) are being slowed by incomplete reporting and inadequate sampling of affected calves and related individuals. Vigilance in investigating neonatal calf problems is paramount.

The first step to investigate calf problems is to establish a clear phenotype definition of the condition. Your local veterinarian can often assist and will accurately diagnose well-known and common calf conditions. Veterinarians are also helpful in accurately communicating phenotypic features of uncommon or emerging diseases, as well as collecting and properly shipping diagnostic or research samples.

Breed association field officers also have knowledge of disorders in the breed and can be a conduit to further diagnostic assistance. The cell phone camera is a huge asset to facilitate disease reporting — often those images and a brief description of interpretation from the ranch serve as a valuable starting point. These images and initial reports help determine sampling needs. Samples used to diagnose and to evaluate environmental or infectious disease vary by the condition presented.

When calves are reported with a phenotypic or epidemiologic feature suggesting a genetic component may be present, we add sampling to enable harvest and banking of DNA for

research. Blood preserved in an EDTA (purple top) blood tube is the easiest sample for scientists to work with as a DNA source.

When affected animals are deceased, an ear may be used to extract DNA for research. The ear cools rapidly at death and is an easy sample to collect. It is important to report disorders when samples are still available. For research, the affected calf, dam, a few half siblings and the sire are sampled for DNA harvest. Samples are saved, and when the analysis suggests further research is justified, genotypic or sequencing studies are pursued.

The diagnostic program at the UNL accepts referrals from breed associations and veterinarians. I encourage breeders to contact their breed associations to report concerns and to access assistance. A local veterinarian, a local diagnostic facility or the UNL program can all initiate investigations when congenital calf disease concerns arise, and they will work together as a team with breed associations to solve these industry-wide problems. **HW**