

Recordati Rare Diseases Partners With the National Urea Cycle Disorders Foundation To Increase Awareness of the Risks of Hyperammonemia

The Check Ammonia Initiative stresses the importance of checking plasma ammonia levels to help protect the brain

LEBANON, NJ (November 29, 2017) -- Recordati Rare Diseases Inc., a biopharmaceutical company committed to providing orphan therapies to underserved rare disease communities in the U.S., today announced a new **Check Ammonia** awareness initiative. The campaign is intended to help healthcare practitioners (HCPs) recognize the signs and symptoms of hyperammonemia in infants, children and adults, act quickly to test ammonia levels, and to call a metabolic geneticist, as high ammonia levels can be toxic and may be the result of a rare metabolic disease.

Created in collaboration with the National Urea Cycle Disorders Foundation (NUCDF), the **Check Ammonia** initiative includes a website, <http://checkammonia.com>, that provides educational and supportive resources for HCPs. The website is designed to help HCPs quickly recognize the signs and symptoms of hyperammonemia, find out when, why, and how to test for ammonia, and to help connect them with metabolic geneticists in their area. This initiative also features a video that addresses hyperammonemia through the personal accounts of caregivers' and patients' experiences, underscoring the importance of early detection and testing for the condition.

Although hyperammonemia is a medical emergency, early detection may be seen as a challenge. It is critical for healthcare practitioners to suspect hyperammonemia and quickly test for the condition—especially with any unexplained alteration in consciousness or encephalopathy.¹ Delayed diagnosis or treatment of hyperammonemia, regardless of cause, can lead to neurological damage and potentially a fatal outcome.²

"Hyperammonemia due to inborn errors of metabolism can strike at any age," said Cynthia Le Mons, Executive Director of NUCDF. "It's our hope that the **Check Ammonia** initiative will be a valuable resource for medical professionals to help them recognize the early signs of hyperammonemia in infants, children, and adults so patients can receive swift intervention to prevent coma and death."

"People affected by rare diseases are our top priority and at the core of everything we do, so we're grateful to have the National Urea Cycle Disorders Foundation working with us on this important disease awareness initiative," said Paul Stickler, Vice President of Commercial Operations at Recordati Rare Diseases. "In infants, children, and adults, hyperammonemia requires early detection and prompt intervention from medical professionals. Our goal is to help physicians quickly identify the condition by recognizing the signs and symptoms of hyperammonemia, checking patient ammonia levels, and consulting with a metabolic geneticist to determine the next steps."

About Hyperammonemia

Hyperammonemia, a metabolic condition characterized by excess ammonia in the blood, can be life-threatening and may affect patients at any age.¹ Clinical signs and symptoms of hyperammonemia are typically neurological in origin, but they can be generally nonspecific and may suggest several diagnostic pathways.¹ The clinical presentation of neonatal hyperammonemia, which can mimic sepsis, includes non-specific symptoms that are mainly neurological in origin.^{3,4} Hyperammonemia in infants (beyond neonates), children, and adults may be more episodic, difficult to recognize and precipitated by catabolic events, protein overload or certain drugs.^{4,5,6} The causes of hyperammonemia are diverse. Some of the more common causes include liver disease, reactions to drugs, hemolytic disease, or gastrointestinal bleeds and urea cycle disorders (UCDs) or other inborn errors of metabolism (IEMs).^{3,7,8}

About Recordati Rare Diseases Inc.

Recordati Rare Diseases Inc. is a biopharmaceutical company committed to providing often overlooked orphan therapies to the underserved rare disease communities of the United States. Recordati Rare Diseases and our sister company, [Orphan Europe](#), are part of the rare disease business within the [Recordati Group](#), a public international pharmaceutical company committed to the research and development of new specialties with a focus on treatments for rare diseases.

Recordati Rare Diseases' mission is to reduce the impact of extremely rare and devastating diseases by providing urgently needed therapies. We work side-by-side with rare disease communities to increase awareness, improve diagnosis and expand the availability of treatments for people with rare diseases.

The company's U.S. corporate headquarters is located in Lebanon, NJ, with global headquarter offices located in Milan, Italy.

For more information, visit <http://www.recordatirarediseases.com> and follow @RecordatiRare on Twitter.

About the National Urea Cycle Disorders Foundation

The [National Urea Cycle Disorders Foundation \(NUCDF\)](#) is dedicated to saving children and adults from the catastrophic effects of UCDs. NUCDF is a leader in the fight to conquer UCD, raising awareness to improve early diagnosis, catalyzing research, and is a vital resource of information and education for families and doctors. NUCDF is a lifeline for affected families seeking guidance and support, providing mentors for newly diagnosed families and networking UCD families together for support.

For more information, visit <http://www.nucdf.org>

References

1. Haberle J. Clinical practice-The management of hyperammonemia. *Eur J Pediatr* (2011) 170:21-34.
2. Auron A, Brophy PD. Hyperammonemia in review: Pathophysiology, diagnosis, and treatment. *Pediatr Nephrol* 2012;27:207-222.
3. Broomfield A, Grunewald S. How to use serum ammonia. *Arch Dis Child Educ Pract Ed* 2012;97:72-77.
4. Summar ML, Tuchman M. Proceedings of a consensus conference for the management of patients with urea cycle disorders. *J Pediatr* 2001;138 (1 Suppl): S6-S10.
5. Summar ML, Barr F, Dawling S, Smith W, Lee B, Singh RH, Rhead WJ, Sniderman King L, Christman BW. Unmasked adult onset urea cycle disorders in the critical care setting. *Crit Care Clin* 2005;21: S1-S8.
6. Haberle J, Boddaert N, Burlina A, Chakrapani A, Dixon M, Huemer M, Karall D, Martinelli D, Sanjurjo Crespo P, Santer R, Servais A, Valayannopoulos V, Lindner M, Rubio V, Dionisi-Vici C. Suggested guidelines for the diagnosis and management of urea cycle disorders. *Orphanet J Rare Dis* 2012;7:32.
7. Orton DJ, Gifford JL, Seiden-Long I, Khan A, de Koning L. Critically high plasma ammonia in an adolescent girl. *Clin Chem* 2016;62(12):1565-1569.
8. Burton BK. Inborn errors of metabolism in infancy: A guide to diagnosis. *Pediatrics* 1998;102(6): E69.

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