Ketotic Hypoglycemia INTERNATIONAL

FIRST ANNUAL VIRTUAL CONFERENCE

IDIOPATHIC KETOTIC HYPOGLYCEMIA

27-28 FEBRUARY 2021

SCIENTIFIC ADVISORY BOARD

Dr. Henrik Christesen
Clinical Professor
H.C. Andersen Children's Hospital

Dr. David Weinstein
Former Director, GSD Program
The University of Connecticut

Dr. Pratik Shah
Pediatric Endocrinologist
The Royal London Children's Hospital

Dr. Terry Derks
Pediatric Metabolic Diseases
Beatrix Children's Hospital

Dr. Paul Thornton
Medical Director
Cook Children's Medical Center

ketotichypoglycemia.org

Click here to register:
www.ketotichypoglycemia.org/conference

WELCOME

We would like to personally welcome each of you to the first annual conference in Ketotic Hypoglycemia International. Due to the pandemic, we are hosting a virtual conference. All of the presentations will be available after the conference for educational purposes.

We are excited to have this opportunity to come together, patients, caregivers, healthcare professionals, experts, pharma- and biotech companies to share current knowledge, ongoing studies, best practices, patient stories, and our collective vision for the future. Our goal with the conference is to unite the families affected by idiopathic ketotic hypoglycemia, the experts, the healthcare professionals, and the industry so we together can take the first step towards an enhanced understanding of idiopathic ketotic hypoglycemia. We have a line-up of presentations made by our scientific advisory board experts and guest speakers, mixed up with patient stories from our KHI-families from around the world. Our speakers will be ready to answer questions after each presentation and in the “Meet the Experts” session on Saturday.

We hope you leave the conference inspired and informed on the latest knowledge in the field of idiopathic ketotic hypoglycemia.

If you are a patient, a caregiver, a family member, a nurse, a dietitian, a medical student, a researcher, a pharma- or biotech company, or something completely different: Your interest is truly our greatest asset today and tomorrow. We could not accomplish what we do without your support.

Do you have a burning question for the experts?
Please e-mail danielle.drachmann@ketotichypoglycemia.com

See you virtually on 27-28 of February 2021

With gratitude,

Danielle Drachmann
Executive Director

Aimée Goodwin
Head of Digital Communication

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SATURDAY 27 FEB

All time slots are in **Copenhagen** time. To find out what the time is in your local time zone, click **here**

08:30pm - 00:30am India (IST)
06:00pm - 10:00pm Russia (MSK)
05:00pm - 09:00pm Africa (CAT)
03:00pm - 07:00pm GMT (U.K.)

04:00pm - 08:00pm CEST (Denmark)
04:00am - 08:00am New Zealand (NZDT)
02:00am - 06:00am Australia (AEDT)
08:00am - 12:00am USA (Mountain Time)

09:00am - 01:00pm USA (CST)
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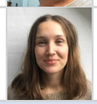
07:00am - 11:00am USA (PT)

Time CEST	Description	Speaker
04:00pm - 04:20pm	**Opening session**	
04:00pm-04:05pm	Welcome	 Danielle Drachmann Executive Director of Ketotic Hypoglycemia International
04:05pm-04:20pm	**Opening session** 04:05pm: The establishment of Ketotic Hypoglycemia International 04:15pm: Introduction to the Board of Directors 04:20pm: KHI Ambassador; Hannah Jordans Story	
04:30pm-06:30pm	**Defining ketotic hypoglycemia**	
04:30pm-04:55pm	A historical perspective of ketotic hypoglycemia *- 10 min. Patient story or/and Q&A*	 Dr. Henrik Thybo Christesen, Odense University Hospital, Denmark
04:55pm-05:15pm	What is hypoglycemia? *- 10 min. Patient story or/and Q&A*	 Dr. Paul Thornton Cook Children's Hospital, US
05:15pm-05:35pm	What are ketones? *- 10 min. Patient story or/and Q&A*	 Dr. Terry Derks Beatrix Children's Hospital, The Netherlands
05:35pm-05:55pm	What is ketotic hypoglycemia? *- 10 min. Patient story or/and Q&A*	 Dr. David Weinstein Passage Bio, US
05:55pm-06:15pm	Symptoms and signs of ketotic hypoglycemia *- 10 min. Patient story or/and Q&A*	 Dr. Pratik Shah The Royal London Children's Hospital, UK
06:15pm-06:30pm	Clinical investigations into ketotic hypoglycemia *- 10 min. Patient story or/and Q&A*	 Dr. Henrik Thybo Christesen, Odense University Hospital, Denmark
06:30pm	15 min. break	
06:45pm-07:30pm		
06:45pm-07:05pm	New genes: Genetic investigations in ketotic hypoglycemia *- 10 min. Patient story or/and Q&A*	 Professor Klaus Brusgaard Department of Clinical Genetics, Odense University Hospital, Denmark
07:05pm-07:30pm	GSDIXa and KH *- 10 min. Patient story or/and Q&A*	 Dr. Anne Benner Odense University Hospital, Denmark
07:30pm-08:00pm	**Closing session**	
07:30pm-07:55pm	Meet the experts: - Your questions answered by the Scientific Advisory Board	 Danielle Drachmann Executive Director of Ketotic Hypoglycemia International
07:55pm-08:00pm	Closing remarks	

SUNDAY 28 FEB

All time slots are in **Copenhagen** time. To find out what the time is in your local time zone, click **here**

08:30pm - 00:30am India (IST)
06:00pm - 10:00pm Russia (MSK)
05:00pm - 09:00pm Africa (CAT)
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04:00pm - 08:00pm CEST (Denmark)
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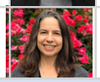
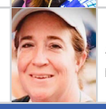
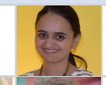
10:00am - 02:00pm USA (EST)
07:00am - 11:00am USA (PT)

Time CEST	Description	Speaker
04:00pm -04:05pm	Welcome	 Danielle Drachmann Executive Director of Ketotic Hypoglycemia International
04:05pm-04:20pm	Collaboration between organizations: KHI & CHI	 Julie Raskin Executive Director at Congenital Hyperinsulinism International
04:20pm-17:10pm	**Management and monitoring**	
04:20pm - 04:45pm	Monitoring KH *- 10 min. Patient story or/and Q&A*	 Dr. Terry Derks Beatrix Children's Hospital, The Netherlands
04:45pm-05:10pm	Prevention and acute treatment of KH *- 10 min. Patient story or/and Q&A*	
05:10pm-06:30pm	**The every-day-life of the KH families**	
05:10pm-05:25pm	Well-being in the KH family *- 10 min. Patient story or/and Q&A*	 Beccie Davis-Yates BSc, PGCE, MEd, AFHE
05:25pm-05:50pm	Eating aversions in children with ketotic hypoglycemia *- 10 min. Patient story or/and Q&A*	 Kathy Ross Metabolic Dietician, Connecticut Children's Medical Center
05:50pm-06:10pm	My life with unexplained ketotic hypoglycemia *- 5 min. Q&A*	 Hannah Jordan KHI ambassador
06:10pm-06:30pm	A mothers story: From surviving to living life to the fullest *- 5 min. Q&A*	 Alicia Jordan Mother and caregiver
06:30pm	15 min. break	
06:45pm-07:55pm	**Ongoing studies**	
06:45pm-07:05pm	Normal values for glucose and ketones *- 5 min. Q&A*	 Dr. Komal Parmar Connecticut Children's Hospital, US
07:05pm-07:25pm	Down Syndrome and ketotic hypoglycemia *- 5 min. Q&A*	  Austin Carrigg US Henrik Thybo Christesen Odense University Hospital, Denmark
07:25pm-07:45pm	Clinical spectrum and genetic basis of ketotic hypoglycemia in children *- 5 min. Q&A*	 Dr. Maria Melikian MD, PhD, Head of the department of the neonatal endocrinopathies in Endocinen Research Center, Moscow, Russia
07:45pm-08:00pm	Comorbidity survey (a patient-initiated project) *- 5 min Q&A*	  Erica Hoffmann BSN, RN Valerie Weaver BSN, RN, TCRN, NRP
08:00pm-08:15pm	**Closing session**	
08:00pm-08:10pm	Next steps for Ketotic Hypoglycemia International	  Jacob Sten Petersen, Chairman of Ketotic Hypoglycemia International Danielle Drachmann Executive Director of Ketotic Hypoglycemia International
08:10pm-08:15pm	Closing remarks	

Virtual Conference I 2021 I 27-28 FEB

IDIOPATHIC KETOTIC HYPOGLYCEMIA

MEET OUR EXPERTS

Ketotic Hypoglycemia International aims to challenge current perceptions and attitudes to expand the recognition that IKH is more than a normal variation and establish an international platform for future research collaboration.
Our mission is supported by leading medical experts within the field of inherited metabolic diseases and pediatric endocrinology united in our Scientific Advisory Board.

Dr. Henrik Christesen

Odense University Hospital, Denmark

Henrik Thybo Christesen is a clinical professor at Hans Christian Andersen Children's Hospital, Odense University Hospital, Denmark and a pediatric endocrinologist with special clinical and research interest in hypoglycemia. He leads the multidiciplinary national team in complex hypoglycemia at the hospital, also receiving international patients. Through international collaboration, his research has led to many published papers in the field, mostly on congenital hyperinsulinism, but also, more recently, on ketotic hypoglycemia. Of more than 110 papers, he has published over 40 papers on glucose disorders and beta cells. He functions as the head of the scientific advisory board in Ketotic Hypoglycemia International.

Dr. Paul Thornton

Cook Children's Hospital, US

Paul Stephen Thornton is a Medical Director, Distinguished Consultant of the Endocrinology/Diabetes Clinic and the Hyperinsulinism Center at Cook Children's Medical Center, in Fort Worth, Texas. His recent awards include the Cook Children's Clinical Scholar Award 2012-2014 and recipient of the 2012 Cook Children's Health Care System Endowed Chair Award. He has coauthored more than a dozen book chapters and over 50 journal articles. Paul is the lead author of the recent Pediatric Endocrine Society's Recommendations and Management of Persistent Hypoglycemia in Neonates, Infants, and Children. Dr. Thornton is also a member of the Pediatric Endocrine Society, Society for Pediatric Research, and The Endocrinology Society.

"Through my work with children with idiopathic ketotic hypoglycemia, it struck me that many of them really have a disease being much more than normal variation. This has prompted us to dive into various genetic explanations and improved management and treatment"

- Dr. Henrik Christesen

Dr. David Weinstein

Passage Bio, US

Following his graduation from Trinity College (CT) and Harvard Medical School, Dr. Weinstein did a residency, chief residency, and fellowship in pediatric endocrinology at Boston Children's Hospital. He subsequently obtained a Masters in Clinical Investigation from Harvard and MIT, and became Director of the Glycogen Storage Disease Program at Boston Children's. In 2005, Dr. Weinstein moved to the University of Florida where he directed the Glycogen Storage Disease Program and became a tenured professor. He and his team moved to the University of Connecticut and Connecticut Children's Medical Center in 2017 to perform gene therapy for GSD. In August 2020, he left the academic world to serve as the medical lead for the GM1 gangliosidosis gene therapy trial at Passage Bio.

Dr. Weinstein has published over 100 manuscripts and 28 textbook chapters in the field of disorders of carbohydrate metabolism. He was named as one of the inaugural Goldwater Scholars in 1989. He is a former Jan Albrecht Award winner from the American Association for the Study of Liver Diseases, and he was the George Sacher Award winner from the Gerontological Society of America. In 2013, Dr. Weinstein was honored with the Order of the Smile international humanitarian award, and he was knighted in Poland as part of this recognition. He has also been inducted in the Rare Disease Research Hall of Fame.

Dr. Terry Derks

Beatrix Children's Hospital, The Netherlands

Terry Derks has been certified as consultant pediatric metabolic diseases and he is appointed as associate professor at the University Medical Center Groningen, University of Groningen, the Netherlands. Since his PhD thesis defense in 2007 (title: "MCAD deficiency: clinical and laboratory studies"), he developed a special interest, both clinically and scientifically, in inherited disorders of glycogen metabolism (i.e. hepatic glycogen storage diseases) and fatty acid oxidation disorders. Dr. Derks has more than 60 international publications, 2 book chapters and is a SSIEM Advisory Council Member. In MetabERN Dr. Derks is a Medical Executive Board member and the Work package leader for the "Virtual Counseling Framework".

Dr. Pratik Shah

The Royal London Children's Hospital, UK

Pratik Shah is a Consultant in Pediatric Endocrinology/Diabetes and Honorary Senior Lecturer at The Royal London Children's Hospital (Barts Health NHS Trust) and Queen Mary University of London (QMUL), London. He did his PhD in the field of Congenital Hyperinsulinism and specializes in children with all forms of glucose disorders and has led the highly specialized service for Congenital Hyperinsulinism. His main research includes understanding molecular basis various forms of non-diabetic hypoglycemia in children and identifying novel therapies in hypoglycemia/hyperinsulinism. He has been a speaker at various international and national meetings and has been primary investigator for various investigational studies and clinical trials.

Ketotic Hypoglycemia INTERNATIONAL

What is ketotic hypoglycemia?

"Ketotic hypoglycemia may be unexplained, or idiopathic (IKH). This is a challenge and should urge for more research"

For the patients

In a normal person, fuel for the brain and the general cell metabolism primarily comes from the burning of sugar deposits (glycogen). When the glycogen stores are depleted, the body will switch to burn fat deposits. The fat burn lead to two fuels for the brain, both glucose (sugar) and ketone bodies. However, ketones in the blood will lead to nausea and eventually vomiting. This will lead to a vicious circle, where you cannot eat or drink sugar-rich items, which again leads to further fat burn and production of ketone bodies.

In a KH-patient, the glycogen stores are somehow insufficient. This leads to decreased fasting tolerance with earlier onset of fat burn and hence ketone bodies. In most patients, the hypoglycemia is relatively mild, and the ketone bodies help to provide fuel to the brain, which prevents loss of consciousness and convulsions. However, in relatively few patients, the condition is more severe, but still without an identified cause despite intense investigations in hormones and cell metabolism. Such patients are said to have "idiopathic" KH, or IKH, which simply means KH without any known cause.

Source: Professor Henrik Christesen, Odense University Hospital, Chair of the Scientific Advisory Board in KHI, Denmark

Illustration by Illulines

For the doctors

Ketotic hypoglycemia can be seen in children because of growth hormone deficiency, cortisol deficiency, metabolic diseases with intact fatty acid consumption, including glycogen storage diseases (glycogenosis; GSD) type 0, III, VI, and IX, or disturbances in transport or metabolism of ketone bodies. When these diagnoses are excluded, ketotic hypoglycemia can be categorized as unexplained or idiopathic (IKH), otherwise known as accelerated starvation.

The primary treatment is dietary prevention with long carbohydrates (e.g. uncooked cornstarch) and frequent meals. In some more severely affected patients, continuous feeding may be needed through a gastrostomy tube.

Emergency treatment constitutes of oral or i.v. glucose, eventually i.m. glucagon, to raise the plasma glucose, which will prevent further lipolysis. However, the ketones can take hours to be eliminated. In more severely affected patients, the ketone production can be significant before hypoglycemia occurs, why recognition of increased ketones above approximately 1.5 mol/L is important, sometimes already when the glucose concentration goes below 3.9 mmol/L (70 mg/dL).

Source: Professor Henrik Christesen, Odense University Hospital, DK

What is this organization about?

”Idiopathic ketotic hypoglycemia is the most frequent state with low blood sugar in preschool kids”

KHI aims to globally unite leading hypoglycemia experts with patients, caregivers, and healthcare providers, and to establish a greater understanding of IKH in the hopes to facilitate improved treatment options and greater quality of life for individuals suffering from IKH.

The organization aims to challenge current perceptions and attitudes to expand the recognition that IKH is more than a normal variation and establish an international platform for future research collaboration. We are committed to support and advocate for those in the KH community through our international KHI Parents Support Group by utilizing the contributions of general medical guidance made by our scientific advisory board, supporting research in the general well-being of KH-families, and furthering research into KH etiology, diagnosis, and management.

Where to find us

IKH-families are united through a variety of social media channels. Ketotic Hypoglycemia International are active on LinkedIn, Instagram, Twitter, and Facebook.
The Facebook Support Group is where the families are most active, and where we are sharing experiences and support each other to navigate the everyday-IKH-rollercoaster ride.

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