

MEET OUR TEAM

Endocrinology

Charles A. Stanley, M.D., *Director,
Congenital Hyperinsulinism Center*

Diva D. De León-Crutchlow, M.D.

Andrea Kelly, M.D.

David Robert Langdon, M.D.

Andrew Palladino, M.D.

Nursing Staff

Lori P. Halaby, M.S.N., C.R.N.P.

Amanda Lee, M.S.N., C.R.N.P.

Ashley Murray, M.S.N., C.R.N.P.

Linda Steinkrauss, M.S.N., C.P.N.P.

Susan A. Becker, R.N., B.S.N.

SURGERY

N. Scott Adzick, M.D.

*Chair, Department of Surgery
Surgeon-in-chief*

PEDIATRIC ANESTHESIOLOGY

Ari Weintraub, M.D.

RADIOLOGY

Lisa J. States, M.D.

Hongming Zhuang, M.D., Ph.D.

Diego Jaramillo, M.D., M.P.H.
Chief, Department of Radiology

PATHOLOGY

Pierre Russo, M.D.

Eduardo Ruchelli, M.D.

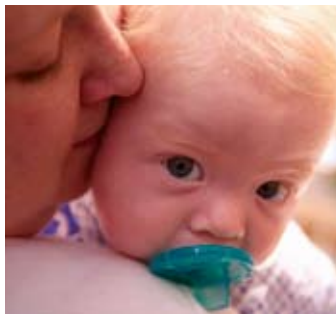
NEONATOLOGY

Jacquelyn Evans, M.D.

Rebecca A. Simmons, M.D.

DEVELOPMENT

Melanie Cohen



*Ty, 6 months, with mom
Denise, is being treated for
hyperinsulinism.*

Hi!

Welcome to the inaugural issue of *HI Hope*, the patient and family newsletter of the Congenital Hyperinsulinism Center at The Children's Hospital of Philadelphia. Through *HI Hope*, we'll help keep you updated on what's new in hyperinsulinism care and research, introduce you to new members of our staff, and provide nutrition tips and other useful advice. We'll publish *HI Hope* four times a year. Should you have questions or ideas for topics,

please e-mail hyperinsulin@email.chop.edu or let us know during a visit to the center. Our patients and families are very important to us. We look forward to staying in touch through *HI Hope*! For further information and links to other resources, please visit our Web site, hyperinsulinism.chop.edu.

Team Players: Staff News

Introducing two new members of the HI staff

Ashley Murray, M.S.N., C.R.N.P., is our newest nurse practitioner. She works as the inpatient nurse practitioner, overseeing the daily lives of patients with HI and other disorders of hypoglycemia. Ashley graduated from Villanova University with a bachelor of science in Nursing and worked as a staff nurse in the Neonatal Intensive Care Unit at Thomas Jefferson University Hospital. She came to CHOP in November of 2008, after completing a master of science in Nursing at Thomas Jefferson University. She enjoys spending time with family and friends, running, reading and traveling.

Andrew Palladino, M.D., came to CHOP in 2006 for his fellowship in Pediatric Endocrinology and has recently become a member of the faculty in the Division of Endocrinology and Diabetes. Dr. Palladino attended Drexel University College of Medicine and completed his pediatric residency at A.I. duPont Hospital for Children and Thomas Jefferson University. During his fellowship, Dr. Palladino's research was focused on describing the molecular mechanisms of various forms of hyperinsulinism. In September, Dr. Palladino assumed the role of inpatient Endocrine Attending Physician, which we hope will improve consistency and continuity of care.

HI Hope: Family Story

In Philadelphia, a Cure

Meghan and Alan of Plymouth, Mass., started 2009 with an unexpected gift. Four days into the new year, their second daughter was born five weeks early. Despite being preterm, Mackenzie weighed 7 pounds, 9 ounces. Mom, Dad and sister Casey were thrilled, but the neonatologist attending the birth began asking questions: Had Meghan had gestational diabetes? Was she diabetic? Was there any history of low blood sugar in her family? The answer was “no” to all.

Shortly after birth, Mackenzie’s blood sugar was 26 mg/dL, much lower than normal. The infant was taken to the Special Care Nursery and given IV dextrose to correct her sugars. Her parents were told that as soon as Mackenzie had five normal blood sugars, she could come out of the Special Care Nursery.

That never happened. At 5 days of age, Mackenzie was transferred to the neonatal intensive care unit of a large university hospital in Boston. Over the next month, she required more and more dextrose to keep her sugars stable. An endocrinologist tested her for hyperinsulinism, and the results came back consistent for the disease. Mackenzie had genetic tests for the congenital form of hyperinsulinism; results would take up to six weeks. Treatment was started, and at 8 weeks of age Mackenzie was discharged.

The next month at home was difficult. Mackenzie needed to be fed every three hours to maintain a blood sugar above 40 mg/dL. She had severe reflux, and if she spit up, her sugar level would drop into the 30s. It was a never-ending cycle.

The results of Mackenzie’s genetic tests came back positive. Mackenzie had a genetic mutation inherited from her father, which was compatible with possible focal congenital hyperinsulinism, meaning it was possible that a small area of her pancreas was affected. Meghan and Alan’s doctor told them: “If this were my child, I would take her to The Children’s Hospital of Philadelphia.”

And so they did.

“The first day I was at CHOP, I was so impressed and relieved,” Meghan remembers. “Clearly they knew what was going on, and what to do, every step of the way. They were on top of their game.”

The HI team scheduled Mackenzie for an ¹⁸F-DOPA PET scan, which revealed a focal lesion on her pancreas. She was then scheduled for surgery to remove a small portion of her pancreas.

“The nurse kept us informed during the surgery, which was great,” her mom recalls. “Afterwards, the surgeon, Dr. Adzick, came out and talked to us.”

Mackenzie spent 22 days in the Hospital. A few days before she was discharged, Mackenzie and her mom were napping when Mackenzie spit up. When Meghan woke up, she called the nurse in a panic, remembering the days when Mackenzie’s sugar levels would plummet as soon as she vomited. The nurse tested Mackenzie’s blood, and her sugar level was normal at 115 mg/dL. “It had never been 115, ever! I was stunned, excited and relieved.”

Mackenzie was discharged at 4 months of age, and returned to Plymouth. Today she is a happy, developmentally appropriate 7-month-old who loves eating and sleeps through the night! She returned to CHOP in August 2009 for her final fasting test, and this confirmed that she is cured.

Childhood is
a *gift* and you
can *give it*.

*For information about giving to the
Hyperinsulinism Program, contact
Melanie Cohen 267-426-6461.*

 The Children’s Hospital of Philadelphia®
GIFT of CHILDHOOD.COM

What's Eating You? Notes on Nutrition

Shiela Falk, R.D., L.D.N., and
Amanda Lee, M.S.N., C.R.N.P.

Parents of infants with HI often worry about what will happen when they transition their child from breast milk or formula to cow's milk at approximately 1 year of age. You may have questions such as, "Will this affect my child's blood sugar?", "How many calories and carbohydrates are in breast milk and formula compared to cow's milk?", and "How often should I be checking my child's blood sugars?"

The nutritional content of cow's milk is very similar to that of breast milk and formula. Eight ounces of breast milk or standard formula contain approximately 160 calories and 17 grams of carbohydrate. Eight ounces of whole cow's milk contain 150 calories and 12 grams of carbohydrate. This should help set your mind at ease! If it helps you feel more comfortable, check blood sugars more often during the transition to cow's milk.

Should you decide to transition your child to soy milk or rice milk, be aware that their nutritional content is very different from that of cow's milk. Eight ounces of soy milk contain 100 calories and 8 grams of carbohydrate. Eight ounces of rice milk contain 120 calories and 24 grams of carbohydrate. Additionally, rice milk contains very little protein and fat to support your growing child's nutritional needs.

Hyperinsulinism Highlighted at Pediatric Endocrine Meetings Around the World

The CHOP HI Center team was well-represented at the recent International Pediatric Endocrine Societies meeting in New York, Sept. 9-12. This was a joint meeting of pediatric endocrine specialists from around the world. Lisa States, M.D., a CHOP radiologist and the primary investigator on the HI Center ¹⁸F-DOPA PET study, presented results and demonstrated potential pitfalls in interpreting images. There were nine talks and eight posters presented by CHOP clinicians, of which two were specifically related to HI. One poster (with our PET results) was recognized as being among the best abstracts submitted to the meeting.

Also in New York, Congenital Hyperinsulinism International (CHI), a parent-initiated nonprofit group, hosted a dinner to foster discussion between conference attendees interested in HI, the CHI board and local families. The dinner was a huge success.

Charles Stanley, M.D., and other conference attendees were leaving New York to attend an International Congress on Congenital Hyperinsulinism being held in Germany the following week.

Hope on the Horizon: Research News

The Congenital Hyperinsulinism Center has numerous promising research studies under way. Here are details of two.

Potential New Medical Therapy

Principal Investigator: Diva De Léon-Crutchlow, M.D.,
Endocrinology

Study Coordinator: Stephanie Givler

We are doing research studies to learn about the effect of a peptide known as exendin-(9-39) on blood sugar levels in children with congenital hyperinsulinism. We are exploring the effect of this peptide to evaluate its potential as a new treatment. One pilot study that is nearing completion involves older children and adults with hyperinsulinism due to mutations in the potassium channel genes (SUR1 and Kir6.2). Two additional studies involving this drug are recruiting younger children and infants now. We hope this research will help us develop new therapies to treat hyperinsulinism. Contact Stephanie Givler at 267-426- 7622 or givler@email.chop.edu to learn more. This research study is supported by the National Institutes of Health and the Katherine and Clifford Goldsmith Philanthropic Fund.

¹⁸F-DOPA PET Imaging

Principal Investigator: Lisa States, M.D., Radiology
Study Coordinator: Susan Becker, R.N., B.S.N.

Infants who need surgery for the treatment of hyperinsulinism that is resistant to medical therapy may qualify for inclusion in our ongoing evaluation of ¹⁸F-DOPA PET imaging to localize a potential focal lesion in the pancreas. While PET scanners are available in many large medical centers, in the United States the drug ¹⁸F-DOPA is only available through a special approval from the FDA. We are collaborating with the Cyclotron Facility of The Hospital of the University of Pennsylvania to make this test available to our infants before surgery.



A Distinguished Ranking

The Children's Hospital of Philadelphia has been ranked first in Diabetes and Endocrine Disorders in U.S. News Media Group's edition of America's Best Children's Hospitals, featured in the August issue of *U.S. News & World Report*. CHOP was ranked No. 1 in more specialties, including Neonatal Care, than any other hospital and is the only hospital that scored in the top three in all 10 of the specialties ranked.

Don't Forget! Reminders for HI Parents

Because of the risks associated with hypoglycemia, we recommend that every child with HI undergo formal developmental assessments at 2 and 5 years of age. Your pediatrician should be able to recommend a developmental pediatrician or specialist for you to see. We would love to hear the results of this testing and how your child is doing!

3/26/09/PA/09_09

THE CONGENITAL HYPERINSULINISM CENTER
The Children's Hospital of Philadelphia
34th Street and Civic Center Boulevard
11 NW Tower, Suite 30
Philadelphia, PA 19104-4399

Non-Profit Organization
U.S. Postage
PAID
Philadelphia, PA
Permit No. 2733

 The Children's Hospital of Philadelphia®

referrals ■ appointments ■ information
215.590.7682 ■ hyperinsulinism.chop.edu