



UNIVERSITY HOSPITAL

Department of Pediatrics
750 East Adams Street
Syracuse, NY 13210

www.upstate.edu/peds/

Nonprofit Org.
US Postage
PAID
Permit No. 110
Syracuse, NY

DEPARTMENT
OF PEDIATRICS
State University of New York
Upstate
Medical
University
UNIVERSITY HOSPITAL
Syracuse, New York

Upstate KidStuff

Volume 4, No. 2

Fall 2006

Calendar

Pediatric Grand Rounds
Wednesdays, 9:15 to 10:30 a.m., Room 6500, University Hospital, Syracuse

October 11
Bassem I. Razzouk, MD, Associate Member, Department of Hematology/Oncology, St. Jude Children's Research Hospital

October 18
Frank A. Oski, MD, Visiting Professor
Alan Lake, MD, Associate Professor of Pediatric, The Johns Hopkins University School of Medicine. The Maryland Pediatric Group, LLC

October 25
Larry Greenburg, MD

A Newsletter for Physicians and other Health Care Providers Interested in Children's Health

FROM THE CHAIR

I hope you had a healthy summer! Things have been very busy in the department, as we start construction for the children's hospital.

Summer brought several new faculty to the department. Drs. Gloria Kennedy (hematology-oncology), Larry Kurlandsky (pulmonology and allergy/immunology), and Greg Liptak (child development) are new subspecialists now in place. Drs. Mary Green, Toby Kressel, and Ellen Schurman are academic generalists in our suburban CareBest practice in Baldwinsville.

As you have probably heard, Upstate Medical University now has a new

president. Former chancellor of Texas Tech University, Dr. David Smith has also joined the department. He is a pediatrician by training, and expects to undertake some clinical work in our general division. Dr. Smith's wife, Dr. Donna Bacchi-Smith, is also a pediatrician. She is trained in public health, and was a very visible advocate for youth tobacco prevention while in Texas. She will be working with us, as well as with the Onondaga County Health Department.

*Thomas R. Welch, MD,
Professor and Chair
Department of Pediatrics*



Architect's rendering of the north/east facade of the University Hospital expansion including the Golisano Children's Hospital which will occupy the top two floors and the tree-house entrance seen at top left.

ePed Direct

This summer, the Department of Pediatrics introduced Golisano Children's Hospital's "ePed Direct" service for referring physicians and other health care providers.

This is a secure, web-based system by which you can send non-emergency questions or comments to pediatric medical and surgical specialists. All that is needed to use the system is an internet connection and email access. When one logs onto the

system for the first time, a brief registration procedure takes place, after which a sign-in name and password are established. After that, the system can be accessed at any time from any place.

Obviously, this system is not designed to address emergencies. But for the types of questions about diagnosis, management, or preventive care which often require a phone call to one of our specialists, ePed Direct will be a great alternative resource.

Questions? Comments? Contact Thomas Welch MD, Department of Pediatrics, 315-464-5451 or welcht@upstate.edu

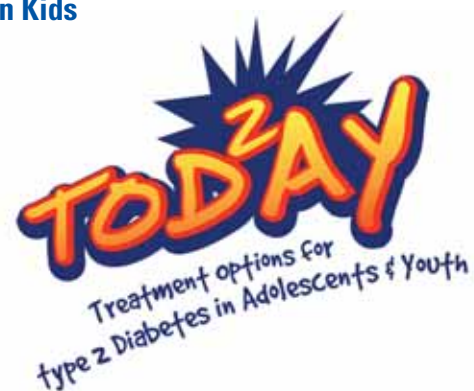
All contents copyright ©2006 SUNY Upstate Medical University

www.upstate.edu/peds/kidstuff.shtml

Update on Today: Treating Type 2 Diabetes in Kids

We are continuing to enroll youth in Treatment Options for Type 2 Diabetes in Adolescents and Youth (TODAY), which is a multi-center clinical research study funded by the National Institutes of Health. It is examining the safety and efficacy of three treatments for type 2 diabetes in the pediatric population. TODAY was initiated due to the increase in type 2 diabetes in children and adolescents, which itself is linked to the rise in obesity and sedentary behaviors in American youth.

TODAY will enroll 750 children, age 10-17, at 13 sites across the United States, and will follow them for two to five years. Enrollment began in May 2004 and continues through April 2007. As of May 2006, 270 children and adolescents with type 2 diabetes have been randomized. All diabetes care, medications and supplies, as well as transportation costs, are provided



without cost to subjects and their families. Dr. Roberto Izquierdo and a team of diabetes educators provide medical care.

As a health care provider, you may have children in your practice with Type 2 diabetes who may be eligible for the TODAY study. If you do, or if you have any questions, please call Ms. Kelly Duncan (Study Coordinator) at 315-464-3878.

Lipid Clinic

The Section of Pediatric Endocrinology is pleased to announce the launch of a program for children with familial hyperlipidemias. Resources for such children and their families in our catchment area have been sparse.

The new Lipid Clinic, staffed by Susan E. Stred, MD, Barbara Lindenmayer, RN, PNP, Lynn G. Horowitch, RN, PNP, and Maryann Russo, RD, CDE, will provide medical nutrition therapy and any indicated medication to those children under 16 years of age who have total fasting cholesterol over 300 mg/dl or total triglyceride over 500 mg/dl.

Lifestyle modification through the primary care office and/or the Healthy Lifestyles Program remains the most appropriate management for the more common pattern (mild elevation of total cholesterol, LDL, and triglyceride, with concomitant low HDL) that accompanies overweight, and the attendant risk for the metabolic syndrome.

Referrals to the Lipid Clinic may be initiated by calling 315-464-6064; preliminary laboratory profiles will be requested prior to scheduling the first appointment.

Faculty Recognitions

Neal Seidberg, MD, assistant professor in our critical care division, was recently awarded the President's Award for Outstanding Contribution of the Year – Hospital, Clinical as well as the Association of Medical Directors of Information Systems Award. In addition to his work as an intensivist, Neal has functioned as the medical director of the hospital's computerized physician order entry (CPOE) process. In this role, Neal was responsible for interfacing with the vendor, training, and helping individual departments "go live" with CPOE. Only about 5 percent of academic medical centers in the country have CPOE, and under Neal's guidance the transition at University Hospital was unusually smooth. With CPOE under his belt, Neal is now helping in the selection of an electronic medical record (EMR) product.

SUNY Upstate Medical University is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

SUNY Upstate Medical University designates this continuing medical education activity for a maximum of 1 category 1 credit toward the AMA Physician's Recognition Award. Each physician should claim only those credits that he/she actually spent in the activity.

Newborn Screening and the Inherited Metabolic Diseases (IMD) Specialty Center

Each year, about 254,000 babies are born in New York State, 130,000 of them in Upstate New York. These infants undergo newborn screening for biochemical and endocrinologic disorders, as well as cystic fibrosis, HIV and hemoglobinopathies. New York expanded its screening program from 11 diseases to 44 diseases in the spring of 2005. There will be one additional test added by the fall of 2006. Of these 44 diseases, 39 are considered inborn errors of metabolism. This includes 5 amino acid disorders, 13 disorders of fatty acid oxidation, 15 organic acidemias, 4 urea cycle disorders, one disorder of carbohydrate metabolism and biotinidase deficiency. Up until now, these infants were referred to either Albany or Rochester for follow-up evaluation and testing.

Presenting "Abby" with MSUD

Abby was a 6-day-old African American female, who had an elevated leucine level on her initial newborn screen. She had been feeding poorly, with some frequent stools and emesis. There were other sick family members with similar symptoms, and Abby was afebrile and gaining weight well after a change to soy formula. Her birth history was unremarkable.

On evaluation for follow-up testing of her newborn screen, she was noted to have a glucose level of 51, with mild acidosis and ketonuria. The blood count was normal but the ammonia was elevated at 177. It was suspected that she was a true positive for maple syrup urine disease and she was hospitalized for evaluation and management. Plasma amino acid quantitation confirmed the diagnosis and Abby was started on a special branched chain free formula. Over the course of her hospitalization she became more alert and began to feed well with good weight gain. She was discharged home on a combination of branched chain free formula and regular infant formula. She will continue to require a highly specialized diet for the rest of her life.

Maple syrup urine disease (MSUD) is an autosomal recessive disorder of branched chain amino acid metabolism. It is due to a deficiency in the activity of the branched-chain ketoacid dehydrogenase multienzyme complex. This causes elevated levels of leucine, isoleucine, valine and their corresponding oxoacids to accumulate. Clinically, infants appear normal at birth and then develop poor feeding and vomiting. They subsequently become lethargic, hypertonic and comatose. Abnormal eye movements and seizures are common. Treatment consists of restricting the intake of the three branched-chain amino acids to amounts essential for growth and no more. The branched-chain levels need to be checked frequently. Patients with MSUD are at risk for further episodes of overwhelming illness and coma that can be fatal or lead to significant neurologic damage. Intercurrent illness or dietary indiscretions may prompt an episode.



IMD Center

In the year 2005, there were 3151 positive newborn screens for the 39 metabolic disorders screened for in the state. 43 percent of these screens were for infants in upstate New York. All these infants were referred to regional centers for further evaluation and treatment but none was seen in Syracuse.

Inborn errors of metabolism are a group of disorders that are life threatening. Infants with a positive screen need to be evaluated further. This means that they will require at least one visit to a metabolic center. If they are true positives, then they will require chronic management. Unfortunately, even with good control there are unavoidable long-term medical complications. Therefore, these children may require care from other pediatric subspecialists as well.

IMD Center named by NYS DOH

The New York State Department of Health has now accredited University Hospital as the newest Inherited Metabolic Diseases Specialty Center. This comprehensive center includes a clinical director, clinical nurse coordinator, metabolic nutritionist, social worker, and genetic counselor.

Our team provides follow-up evaluation and testing on infants with a positive newborn screen for any of the 39 tested metabolic disorders. If an infant screens positive, the Newborn Screen Follow-up program in Albany will notify our center and the primary care physician.

We are happy to evaluate the infant and arrange for confirmatory testing. Our IMD program is based in the Center for Neurodevelopmental Pediatrics at 550 Harrison Street in Syracuse. It will soon be moving to the Physician's Office Building on Irving Ave. We can be reached by calling 315-464-6395. Appointments can be scheduled by calling 1-866-543-KIDS.

We will continue to provide chronic care for those individuals with a confirmed metabolic disorder through our IMD center where they can be seen by our nutritionist and social worker. These children and adults often require specialized formula, medical food products, and other amino acid supplements. In addition, we have collaborated with maternal fetal medicine in order to service the women with metabolic disorders during pregnancy.

The newest test to join the NY State screening panel will be for Krabbe disease. Infants who screen positive will be evaluated at the IMD center and confirmatory enzyme testing for galactosylceramide β -galactosidase will be sent.

Once we do, a more complete article in *KidStuff* will describe the system and how it can be accessed.