



Perinatal Gazette

Newsletter of the Regional Perinatal Center
Westchester Medical Center



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Pediatric Fall Grand Rounds Schedule 2001 (Cedarwood Hall at WMC Campus)

- September 12-** "Updates in Stereotactic Radiosurgery"
Dr. Deborah Benzel
- September 19-** "Update on Congenital Heart Disease Life after
the Arterial Switch Procedure" Louise Danzig, PNP
- September 26-** "Reflex Sympathetic Dystrophy in Pediatrics "
Dr. Eric Small
- October 3** -"Hand Guns in the House?" Dr. Cathy Falvo
- October 10** - CPC/M&M Dr. Woolf and Residents
- October 17** -"Legal Survival Skills for Pediatricians in 2001"
Barbara DeCesare RN,J.D.,MPH
- October 24** -"Epilepsy Monitoring in Children" Dr. Vent Raman
- October 31** - Infectious Disease – speaker to be announced
- November 7** -Topic to be announced Frank Hallenbeck/Abbott
- November 14** -"Bone and Soft Tissue Infections:Diagnostic
Problems" Dr. Lorry Rubin
- November 21** – Epidemiology in Peds Dr. MarisaMontecalvo

For directions and/or questions please call Jan at (914) 594-4020

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What's New in Newborn Screening?

Newborn screening has been with us since 1965 in New York State with the introduction of screening for Phenylketonuria (PKU). Since then over 10 million children have been tested and the screening panel of diseases tested has increased to 8. In addition to Phenylketonuria, New York State also screens for Biotinidase Deficiency, Galactosemia, HIV, Homocystinuria, Hypothyroidism, Maple Syrup Urine Disease, and Sickle Cell Disease.

Next year the panel of diseases screened for will increase dramatically. The Newborn Screening Laboratory at the Wadsworth Center in Albany is being upgraded as I write with new equipment and personnel in preparation for the new screening initiatives .

Screening will be introduced initially for Congenital Adrenal Hyperplasia, Cystic Fibrosis and Medium Chain AcylCoA Dehydrogenase (MCAD) Deficiency. Shortly following the introduction of these tests, screening will be expanded for additional fatty oxidation disorders and organic acidemias, using the same tandem mass spectrometry technology for MCAD Deficiency testing.

MCAD Deficiency is the most common defect of fatty acid oxidation; the enzyme is in the breakdown pathway of fats, specifically beta-oxidation. As a result patients with the condition are prone to hypoglycemia with fasting. A typical presentation would be for an otherwise healthy infant to develop an intercurrent illness and stop eating, The patient could then be at risk for developing hypoglycemia. A severe episode can result in death, and unfortunately about one third of patients who present with this condition die during the first episode. Other patients may develop severe neurological dysfunction following the episode. However, once a patient is identified and the correct precautions taken, then there is really no risk to the patient. Treatment revolves around the prevention of fasting, with frequent feed and the use of cornstarch at night, which prolongs glycemic control. With intercurrent illnesses, patients may need to be admitted for intravenous dextrose.

The Tandem Mass Spectrometer (TMS or MSMS) is a very sophisticated instrument that allows for the simultaneous measurement of multiple metabolites. Depending on the set up for the machine, the TMS can detect over 30 indicator metabolites on one test. *Continue on page 3*



**HUDSON VALLEY
REGIONAL INFANT APNEA
CENTER** call (914) 493-7585
by Dr. Nikhil Amin, Division of Pediatric Pulmonology

The Hudson Valley Regional Infant Apnea Center is a joint effort of the Children's Hospital at Westchester Center and its affiliated medical university, New York Medical College, located in Valhalla, NY. Designated by New York State as an infant apnea center, the staff is dedicated to state-of-the-art patient care, research and education; and serves as a resource for both healthcare providers and the community throughout the Hudson Valley.

Frequently Asked Questions about our Apnea Center:

1. **What is significant apnea?** Apnea simply means a pause in breathing. All infants have brief pauses or apnea and they are usually completely normal. However, some infants have longer pauses and these may be serious and require immediate attention. Generally, we consider apnea significant only if the pause results in adverse physiological effects: bradycardia, desaturation, etc. The actual length of the apnea pause is not as important as how the infant "handles" the event.
2. **What is an "ALTE"?** ALTE means Apparent Life Threatening Event. Infants are occasionally found blue or dusky, limp or stiff and appear not to be breathing and may require resuscitation although, they may seem completely normal within few minutes. Nothing is more frightening to a parent or caregivers than infant apnea or an ALTE. There are many possible causes for apnea or ALTE and it is vital that complete evaluation is performed. Our multidisciplinary team has special expertise in the evaluation and management of these episodes, so parents can have confidence that everything possible will be done to minimize any chance of a recurrence.
3. **What roles does the Apnea Center staff play in neonatal care?**
 - **Inpatient consultation** – The apnea center physicians are frequently consulted by neonatologists and pediatricians to assist in the management of infants with persistent apnea.
 - **Interpretation of sleep studies** - Our physicians are trained in the interpretation of polysomnography and a variety of sleep studies, varying from 2 to 16 channels of physiologic data. We interpret studies done in over 20 nurseries and neonatal units.
 - **Management of medications for apnea of prematurity** - Our physicians have tremendous experience in assisting primary care providers in the safe dosage-adjustment and weaning of respiratory stimulants such as caffeine or theophylline.
 - **Management of home infant apnea monitors** – Our physicians are following over 100 infants on apnea monitors at any one time. We are available 24 hours a day, 7 days a week, to evaluate unexpected alarms, minimize false alarms, and reassure families and physicians so that monitoring is helpful and discontinued appropriately.
4. **Where is The Hudson Valley Regional Apnea Center?**

The Center's main clinical facility is now located at
19 Bradhurst Avenue, Hawthorne, New York.
In addition, the center physicians now follow infants with apnea in 5 satellite offices throughout the Hudson Valley
5. **How can pediatricians and patients contact the apnea center?**

Simply call (914) 493-7585. A physician is available 24 hours a day, 7 days a week.
6. **Who are the current members of the Infant Apnea Center staff?**
 - Nikhil Amin, MD Director*
 - Joseph T. Boyer, MD*
 - Allen J. Dozor, MD*
 - Suzette Gjonaj, M.*
 - Cathy Kim, MD*
 - Sankaran Krishnan, MD*
 - Diana B. Lowenthal, MD*
 - Madelint Heydendael, R.N; MSW Apnea Center Nurse*
 - Richard Griffin, MS., Pulmonary Function Laboratory Technician*
 - Michelle McNamara, Center Coordinator*

Continued from page 1

The TMS take about a minute to complete a sample analysis on one patient. The process is automatable, so nearly 1000 samples can be measured on a daily on one machine.

Screening for Congenital Adrenal Hyperplasia (CAH) will detect the salt-losing and virilizing form of 21-Hydroxylase Deficiency, the most common form of CAH. The milder late onset form of the condition will not be detected consistently during the screening process. The screening test measures the level of 17-hydroxylase from the heel stick sample.

Cystic Fibrosis (CF) is the most common autosomal recessive condition affecting the Caucasian population. It is characterized by the development of persistent lung infection and pancreatic exocrine deficiency. It is a progressive condition with a median life span of about 30 years. Early detection of patients allows for earlier therapeutic intervention with nutrition augmentation and initiation of respiratory therapy. The early detection should lead to slower progression of the condition with a resultant expectation for improved quality of life and life span. Screening for CF is done by measurement of immune reactive trypsinogen (IRT) levels. The top 5% of levels will have secondary mutation analysis. Samples with 2 known pathological mutations in the cystic fibrosis transmembrane receptor (CFTR) will be referred to their Regional CF Center. Patients with one detected mutation will be referred for sweat testing; this will lead to the identification of either patient who will be referred to the CF Center of to carriers, who will be referred to for genetic counseling. As it is only feasible to test for only the most common CFTR mutations, therefore it is possible for a patient to have a high IRT level and still have no detectable CFTR mutation. Patients with the top 0.2% of IRT results will also be referred for sweat testing.

In New York State there are designated centers for the diagnosis and management of patients referred from the Newborn Screening Program in Albany. In the Lower Hudson Valley Westchester Medical Center is the referral center for all patients with a suspected inherited metabolic disease. The Inherited Metabolic Disease Center (IMDC) at Westchester Medical Center is one of 7 IMDC's in New York State. The IMDC and similar Center's have the responsibility for seeing all patients referred to them in a timely manner, usually within hours of being notified. They use all available means to locate and evaluate any patient with a potential metabolic disorder. Similar situations exist for all the other conditions that are detected by the Newborn Screening Program.

These new developments in Newborn Screening are further acknowledgment of the importance of prevention and presymptomatic disease detection. With our increasing knowledge of the genetic etiology and pathogenesis of disease processes the logical progression to Newborn Screening is natural. Developing technology will make it feasible to screen for many more diseases in the near future. Whether screening should be expanded further will depend on the acceptance and integration of these current initiatives.

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“Clinical Notes”

As part of our plan of Quality Improvement for compliance with HIV testing, documentation and follow up, we will be:

- Putting together a letter notifying all referring facilities of required acceptable documentation regarding the HIV testing status of the mother/infant prior to transport to WMC.
- Develop a monitoring tool based on information needed and monitor monthly for one quarter.
- Review results at Joint Conference monthly and establish future monitoring needs based on results.
- STAT flight will be notified of change in practice and they will monitor and report results monthly for at least one quarter.
- STAT flight will be monitoring their compliance as well as the availability of the information from the referring institution.

We seek the help of all of our transfer affiliates in optimizing patient services in this area.

New Children's Hospital Faculty:

Dr. Kiran Vohra, is joining the Division of Neonatology and will be based in the Neonatal ICU at Sound Shore Medical Center In New Rochelle as of August 1st. Dr. Vohra completed his pediatric residency at Albert Einstein College of Medicine. Dr.Vohra completed his pediatric neonatology fellowship at Mount Zion Hospital and Medical Center, Cardiovascular Research Institute at the University of California, San Francisco. Dr. Vohra has worked 13 years at Interfaith Medical Center in Brooklyn, New York, and most recently worked as the Director of Neonatal Services at Flushing Hospital Medical Center.

Dr. Pranav Patel is joining us as of July 1st as The first Post-Doctoral Scholar in Perinatal Medicine in the Neonatal ICU. His studies will involve gut maturation. Dr. Patel recently completed his pediatric residency here at Westchester Medical Center/ New York Medical College.

Dr. Sonya Strassberg is joining us as of July 1st as a NICU fellow. Dr. Strassberg recently went to University of Medicine and Dentistry of New Jersey-New Jersey Medical School and completed her pediatric residency at Atlantic Health Systems in Morristown, New Jersey.

March of Dimes "Walk America" 2001 Update

On Sunday April 29, 2001 seven representatives from Westchester Medical Center (AKA "The Westies") gathered at Saxon Woods Park in White Plains to participate in Walk America 2001. The goal of this event was to raise money for the March of Dimes and support their mission of decreasing infant morbidity and mortality. It was a glorious morning with early temperatures in the 40's and reaching mid-70's by late morning. The enthusiasm and energy was very apparent as all the walkers prepared for the 8-mile trek that was before them. It was a great opportunity to socialize, exercise and support a good cause. The participants included: Jen Stavropoulos, Martin Leis, Jackie Questo, Patti Simon, Lissette Paonessa, Eileen Margenat and John Brady. The collective effort of all the walkers and those that donated money by purchasing baby booties, sneakers or stars in the cafeteria on "Hero Day", as well as a matching corporate sponsorship donation yielded \$3,577,00! The overall amount of money collected at the Westchester site was about \$650,000 and they are still counting! Thank you to everyone who participated. We will be forming a committee next year to promote this wonderful event. If you are interested in participating on this committee, please contact me. Thanks again to all the "Heroes" that made this day a great success!

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We are interested in providing you with a newsletter that is relevant and of interest to you. Please contact us with perinatal topics you would like to see addressed.

For a copy of our newsletter or to be placed on our mailing list contact us by phone or e-mail.



"The Westies"

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