

PediNeuroNEWS

Diagnosing and treating children with autism, epilepsy and other neurological disorders.

VOLUME II

DIETARY TREATMENT FOR EPILEPSY HELPS YOUNG PATIENT

Lynzi Janae Rideout started having seizures when she was just ten months old. To stop them her parents tried five different medications before turning to a Vagal Nerve Stimulator when she was four.

It was a relief when the epilepsy was under control, but there was another concern. Lynzi had gained an alarming amount of weight from the medications. Her weight made it difficult for her to run and play like other children. In addition, she had high cholesterol.

The physicians at Boston Medical Center thought the solution might be a strict diet that they use successfully to treat epilepsy.

“We are concerned about every aspect of our patients’ lives, not just their illness,” said Laurie Douglass, M.D. “We also want to ensure that we are offering them the best treatment options, whether that is surgery, medication or a change in eating habits.”

Dietary changes were first used to treat epilepsy in the early 1900s with the introduction of the Ketogenic diet. The strict meal plan is high in fat and low in carbohydrates and includes just enough protein for growth. For a period of time, the Ketogenic diet fell out of use as new epilepsy medications were developed in the 1970s.

In recent years, dietary therapy has regained attention as it has become clear that medications are only part of the solution.

“Once a person has failed two medications, the chance that another will help is just seven percent,” Douglass said. “Although many new medications have come along, that number hasn’t changed.”



The Ketogenic diet has the strongest response in terms of controlling seizures, but physicians at Boston Medical Center have found that a less restrictive low-glycemic diet, which was prescribed for Lynzi, is better for some patients. Although somewhat less effective in controlling seizures, it is more flexible and easier to follow.

On the Ketogenic diet, patients must carefully count calories and weigh foods. Much of the food must be homemade and very few vegetables or fruits are allowed. Patients must stay in the hospital during the first week of the diet to allow physicians to monitor their bodies’ reaction.

In contrast, patients on the low-glycemic diet can eat large amounts of green vegetables along with some fruits and healthy complex carbohydrates. Only about 60 percent of a patient’s calories come from fat compared to 90 percent for the Ketogenic diet. Servings are based on portion size.

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With a successful Spring behind us, it’s fitting that this issue of PediNeuroNEWS emphasize our areas of growth that enhance the exceptional care we offer our patients. Our emphasis on epilepsy care is underscored by features on dietary treatment options, our unique Demo Kitchen, and collaboration with a pediatric Chef-dietitian, Deidra Dexter-Hine. We proudly highlight Dr. Rinat Jonas, a gifted pediatric epileptologist as well as the opening of a newly constructed Pediatric Sleep Clinic and an expanded pediatric sleep program featuring Dr. William DeBassio. We are also fortunate to collaborate with Dr. Mark Korson, a world-class specialist in metabolic disorders from the Floating Hospital for Children at Tufts Medical Center. Finally, we report new ground resulting from collaborative research work in the area of cerebral palsy among preterm infants. May your summer garden be as bright and lush. Happy reading.

Sincerely,

Karl Kuban, M.D.
Division Chief

DIETARY TREATMENT FOR EPILEPSY HELPS YOUNG PATIENT

(Continued from page 1.)

For either diet, the chef-dietitian at BMC works closely with patients and their families to find meals and snacks that are appealing. Patients' plans are drafted individually to meet their needs and tastes.

"It's not easy to say no to treats at a friend's house or to have to measure everything we eat, but the results are astonishing," her

mother, Emily Rideout, said of the complete diet overhaul. "This diet has really made a tremendous improvement in our lives."

Lynzi lost 26 pounds, and her weight and cholesterol are normal for her age. Before she lost the weight, she was winded easily and was unable to keep up with her peers. Lynzi is now playing soccer and running

around like other children her age. She has been seizure-free for more than two years. Physicians have even been able to reduce her dose of antiseizure medication. ■

BMC PEDIATRIC NEUROLOGY DEPARTMENT LAUNCHES PEDIATRIC SLEEP CLINIC AND EXPANDS SLEEP LAB

A good night's sleep is crucial to the health and development of growing children. Without restful sleep, children may do poorly in school or even be sick more often and have trouble fighting off colds. Sleep issues pose serious problems for children with conditions such as epilepsy, congestive heart failure, ADHD, obesity or sickle cell anemia, often contributing to the worsening of these disorders.

Many medical problems may disrupt crucial night rest, or poor sleep may worsen the conditions.

In order to ensure that these children are diagnosed effectively and treated comprehensively, Boston Medical Center's Pediatric Neurology Department has established the Pediatric Sleep Clinic and has expanded the hospital sleep laboratory to better accommodate young patients.

"We recognize that evaluating a child's sleep pattern can be a critical part of an overall diagnosis and treatment plan," said Director of Pediatric Sleep Services William DeBassio, Ph.D., M.D. "At BMC, physicians from various specialties work together to better understand what is happening with our young patients and how best to help them."

It is difficult to assess the magnitude and prevalence of sleep disorders since so many go unreported. The likelihood of having or developing sleep problems increases with certain diseases. Some estimate up to 90 percent of children with learning disabilities may have a sleep disorder. The numbers are almost as high for children with ADHD or cerebral palsy.

Children may be referred to BMC's clinic by physicians at Boston Medical Center or associated clinics or by their own private physicians. A few indications for referral might include daytime sleepiness, difficulty initiating sleep, insomnia, snoring, apnea, sleep walking or talking, night terrors, nightmares or leg pains disrupting sleep.

Sleep specialists at the Pediatric Sleep Clinic can help determine what the sleep problems are, as well as examine whether sleep issues are making other medical problems worse or if other problems are interfering with sleep. Some children who aren't getting enough sleep may display behaviors similar to ADHD, for instance.

The specialists work with patients' primary physicians to develop a treatment plan. Some sleep problems can be eliminated by a change in routine. In other cases, doctors may prescribe a sleep aid to help a child fall asleep at night or a stimulant to help a child stay awake during the day.

For some issues, children are referred to the Sleep Laboratory for an overnight sleep evaluation called a "polysomnogram" to diagnose neurologic, breathing or movement disorders.

Armed with this information, doctors can determine, among other things, whether a patient has obstructive sleep apnea, which may require surgery, or restless leg syndrome, which may be treated with medication, improved diet or exercise.

During the polysomnogram, monitors are used to measure brainwaves, heart rate and eye movements. Sensors placed by the nose and mouth measure airflow, those around



the chest and abdomen measure breathing movements, and a sensor on the finger measures oxygen levels.

BMC has added two rooms specifically designed for polysomnography with children and has specialized technicians who make the experience less stressful. The rooms are large enough to allow parents to stay with their children, and the décor is inviting to the pediatric set.

"We recognize the importance of creating an environment that is designed with children in mind," DeBassio said. "This expansion and modification of the Sleep Laboratory is an important way to welcome families."

Boston Medical Center is a national leader in the relatively young field of pediatric sleep disorders and has recently established a training program to improve other physicians' knowledge about the frequency and significance of these disorders. It is clear that millions of children suffer from significant sleep disorders ranging from insomnia to sleep apnea. The new clinic delivers enhanced treatment for children of all ages as well as the training of doctors in this crucial field. ■

GETTING THE METABOLIC WORD OUT



Mark Korson, M.D., director of the Metabolism Clinic at Tufts Medical Center.

Metabolic disorders are chemical imbalances that interfere with the body's ability to convert food into fuel. These disorders, often inherited, can have severe effects such as mental retardation, seizures and even sudden death.

Early intervention can prevent or alleviate many of these problems, but they too often go unrecognized due to a shortage of physicians trained in the area of metabolic genetics.

Furthermore, few trainees are coming into this specialty.

Boston Medical Center has joined an innovative multi-institutional program created to address these issues through training and promoting awareness of the disorders and symptoms. The Metabolic Outreach Service (MOS) was initiated by Mark Korson, M.D., director of the Metabolism Clinic at Tufts Medical Center's Floating Hospital for Children.

"It is important that nonmetabolic physicians participate more in the diagnosis and management of this patient population," Korson said. "Our goal is to encourage physicians to think more about how these disorders may be affecting their patients."

Through the program, Korson visits six regional medical centers on a regular basis to provide educational and consultative services. The MOS program includes practical, case-based workshops, opportunities for patients and their family members to share their experiences with medical audiences and

consultative assistance to hospital staff around cases in which a metabolic cause is being considered. The effort represents the largest and most comprehensive educational service of its kind in the area of metabolic disease.

"This unique collaboration allows us to share crucial information and find better ways to diagnose and care for our patients," said Karl Kuban, M.D., Boston Medical Center's chief of Pediatric Neurology.

The service is intended to change clinical practice, enabling clinicians to "think metabolic," Korson said. He also hopes the program will encourage medical students and young physicians to consider working with metabolic patients.

The MOS began at Boston Medical Center last November. It is sponsored by Genzyme Therapeutics, Shire Human Genetic Therapies, Ucylyd Pharma, Sigma-Tau Inc., Actelion Pharmaceuticals, Biomarin Pharmaceutical Inc. and the Baby Lorenzo Gregory Scavio Fund. ■

DEMO KITCHEN HELPS FAMILIES DEVELOP MEAL PLAN

At Boston Medical Center, patients and their families now have a special opportunity to learn firsthand about dietary therapies used to treat seizures and epilepsy such as the Ketogenic diet, the Atkins diet, and the low-glycemic diet—in the hospital's unique demonstration kitchen. Chef-dietitian Deidra Dexter-Hine invites parents and children into the Demo Kitchen to prepare and sample a few recipes that might be prescribed for them.

"We felt that we had to do more than just hand patients and their families a stack of recipes and a list of dietary restrictions," said Laurie Douglass, M.D. "They will

have more success in following the diet if they can see how things should be made and cook for themselves."

The tidy kitchen looks like a set for a television show with mirrors arranged over cooking spaces. All the comforts of home make it as close to real life as possible.

Before putting patients on the Ketogenic diet or a variation of it, dietitians meet with families to discuss the food restrictions and to get a sense of how well they will be able to adhere to the changes. When diet is used to treat seizures, cheating may be a consequence. Consuming a small amount of the

wrong carbohydrate can lead to loss of seizure control. Dietitians work with parents to develop a meal plan based on the diet's requirements and the child's eating preferences. Parents at BMC can turn to the culinary expertise of Dexter-Hine to develop creative kid-friendly recipes.

"We want to make sure that families have all the essential information they need to develop a meal plan that they can follow. They are more likely to be successful if they can adapt the diet to their own tastes and lifestyles," said Dexter-Hine. ■

INTRODUCING RINAT JONAS, M.D., PEDIATRIC EPILEPTOLOGIST

Epileptologist Rinat Jonas, M.D., is fascinated by what is known and unknown about the brain and epilepsy.

Jonas is the third epilepsy specialist and the newest member of the team that comprises the Division of Pediatric Neurology at Boston Medical Center. Her focus on the management of intractable seizure disorders, including evaluation for possible seizure surgery, complements her expert skills in interpreting EEGs and other electrophysiological studies, including videotelemetry and electrocorticography (putting EEG electrodes directly on the brain).

She became interested in the mysteries of the brain as a pediatrics trainee in Israel, where she encountered a variety of challenging neurological disorders. “I realized how much of brain development is still unknown,” Jonas said. “It was this realization that served as a strong impetus for me to pursue a career in pediatric neurology.”

Jonas moved to Los Angeles in 1999 and completed her three years of pediatric neu-

rology residency at UCLA, followed by two years of fellowship training in pediatric epilepsy and clinical neurophysiology at UCLA, which has a major pediatric epilepsy center. While at UCLA, Jonas strengthened her commitment to the management of patients with complex seizure disorders, and gained an added expertise in the evaluation of children for possible surgical treatment of uncontrolled epilepsy.

“I have seen a large number of children representing the full spectrum of the childhood epilepsies,” she said. “As I learned, much can be offered to ease the burden of seizures using both pharmacological and nonpharmacological management. On the other hand, a great deal is still unknown, and continued intensive research in this field is critical.”

Her research has advanced the quality of care possible for children with infantile spasms and following traumatic brain injury. Jonas studied the ability of the brain to repair itself (neuroplasticity) and evaluated



Rinat Jonas, M.D., experienced epileptologist and physician-scientist.

language development in children who underwent removal of part of the brain as treatment for uncontrolled epilepsy. Additionally, Jonas has been a contributor to multiple clinical studies that evaluate the effectiveness of antiepileptic drugs.

“BMC is very fortunate to have a physician with such extensive expertise who also has a wonderful bedside manner and a remarkable ability to relate to children and families,” remarked Dr. Karl Kuban, the Chief of the Division of Pediatric Neurology. “We are proud to introduce her to the community we serve.” ■

BMC NEUROLOGIST DEVELOPS ALGORITHM TO CATEGORIZE CEREBRAL PALSY FINDING BETTER WAYS TO DIAGNOSE A COMPLEX DISORDER

Cerebral palsy (CP), a complex collection of nonprogressive muscular disorders caused by brain dysfunction, presents a particular challenge to physicians and researchers because it may be difficult to diagnose and classify for some nonspecialized clinicians. Even experienced physicians might disagree as to whether a patient has a disorder and what type it is.

The lack of a precise diagnosis may make it difficult to give parents an accurate outlook for their child. It also complicates the process of trying to identify specific risk factors associated with the various types of cerebral palsy.

The chronic, nonprogressive condition was first identified more than 160 years ago. It is caused by damage to one or more specific areas of the brain, usually during development, and affects body movement and muscle coordination.

Physicians at Boston Medical Center, led by Karl Kuban, M.D., Chief of Pediatric Neurology, in collaboration with 14 other medical centers around the country, are working on ways to improve current practices and pave the way for innovative research.

“We recognize the challenges that this collection of disorders represents and want

to make it less complicated for physicians, while helping parents understand more about what their child may face,” he said.

Dr. Kuban developed a video program that guides physicians through performance of various aspects of a neurological exam in order to show physicians how to elicit findings.

Following on that work, Kuban led a group of researchers in developing a computerized algorithm that helps accurately diagnose three main types of CP. Physicians enter data about the patient and the program then categorizes the disorder.

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BMC NEUROLOGIST DEVELOPS ALGORITHM TO CATEGORIZE CEREBRAL PALSY, FINDING BETTER WAYS TO DIAGNOSE A COMPLEX DISORDER

(Continued from page 4.)

The results, along with other findings about the prevalence of CP and related complications were published recently in the *Journal of Pediatrics*. The algorithm models the way a seasoned pediatric neurology clinician might identify and classify CP.

Only two in 1,000 children are born with cerebral palsy, but infants born before 28 weeks' gestation have a 50-fold elevated risk compared to babies born at full term. In some children, the disorder is severe,

involving both arms and legs, while others have less impairment. CP may affect just one side of the body or only the arms or legs.

All 1,056 children in Kuban's study were born before 28 weeks' gestation. Of them, 11.4 percent had CP. Of those, 31 percent had diparesis, CP involving one or both legs; 17 percent had hemiparesis, CP on one side of the body; and 52 percent had quadriparesis, CP involving a combination of arms and legs. The study also showed

that children with quadriparesis had higher motor dysfunction and more co-morbidities, such as cognitive disabilities.

"This study shows how commonly the various disorders are seen and how often they are associated with other problems," Kuban said. "This is an important step in understanding the disorders and researching their causes. Once we know more about how they develop, we can find better ways to treat and possibly prevent them." ■



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