Le Bonheur Children's Hospital
Methodist Healthcare Family

Referrals: 888-890-0818
www.lebonheur.org/neuroscience

Brain Waves

Neuroscience Institute

Fall 2010

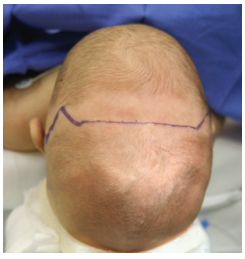
Craniofacial team takes multi-disciplinary approach

Sagittal synostosis case study: Surgeons relieve sutures, reshape skull

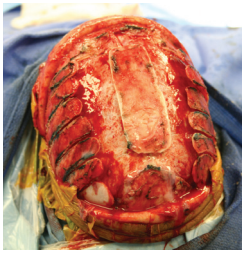
Shortly after Gabriel LaFountaine was born, his parents noticed "weird movements" from their baby boy. They caught him rolling his eyes and clinching his fists. And then, there was the shape of his skull.

The LaFountaines would later learn Gabriel suffered from sagittal synostosis, a premature closure of the skull's sagittal suture, which results in an abnormally long, narrow head, termed scaphocephaly.

After months of looking for the right surgeon, the LaFountaines were referred to Pediatric Neurosurgeon Rick Boop, MD, and Plastic Surgeon Robert Wallace, MD—part of the craniofacial team at Le Bonheur Children's Hospital.



Prone-facing sagittal synostosis before surgery



Prone-facing sagittal synostosis after surgery

The pair performed a cranioplasty on 8-month-old Gabriel this past fall. "I couldn't ask for a better team," said Gabriel's father, Christopher.

Boop and Wallace are part of a multi-disciplinary craniofacial team at Le Bonheur Children's that includes neurosurgeons, plastic surgeons, dentists, orthodontists, therapists and other specialists. The craniofacial program also carries a pediatric craniofacial plastic surgical fellowship program.

Rick Boop, MD, left, and Robert Wallace, MD, right, perform a cranioplasty on 8-month-old Gabriel LaFountaine.

Le Bonheur's craniofacial team specializes in a variety of craniofacial anomalies – including cleft palate, craniosynostosis, congenital mandibular and maxillary deformities, as well as other craniofacial syndromes.

Eight-month-old Gabriel suffered from sagittal synostosis, the most common type of craniosynostosis. Sagittal synostosis occurs in one in 1,000 live births and carries a significant male predominance, according to Boop. The anomaly causes a characteristically abnormal shape of the skull and, in most instances, occurs gradually. The cause is often unknown, but sagittal synostosis typically runs in the male side of a family, suggesting some genetic predisposition. Secondary synostosis may occur following placement of a ventricular shunt for the treatment of severe hydrocephalus.

Treatment is primarily surgical, and the surgical procedure varies depending on the surgeon, age of the patient and severity of the synostosis. For Gabriel, Boop and Wallace were able to surgically relieve fused sutures, so that, through unimpeded growth of the brain, the skull could attain a normal shape. In such instances, the neurosurgeons make surgical cuts in the abnormally shaped skull, releasing the bone where it has grown together prematurely. The plastic surgeons then reshape the skull,

continued on page 3

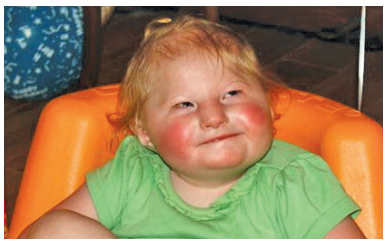
A pediatric partner with the University of Tennessee Health Science Center/College of Medicine and St. Jude Children's Research Hospital

Anti-seizure Ketogenic Diet Brings Hope to Families

Dedicated ketogenic dietitian provides continuity of care

When seizure medicines failed to treat 2-year-old Helen Weber's infantile spasms, her neurologist suggested that her parents look at other options.

Because the two medications Helen had been taking weren't working, Eric and Annie Weber weren't eager to add another. So when a neurologist suggested they consider the ketogenic diet to treat seizures, the Webers weighed their options.



The ketogenic diet has helped relieved 3-year-old Helen Weber's seizures.

Well versed in epilepsy treatment options, the Webers knew the strict, high-fat ketogenic diet could be successful in treating seizures, especially infantile spasms. They also knew it is a lot of work and would require a great commitment for their family. With eyes wide open, they decided to try the diet.

A year later, neurologists and the ketogenic dietitian at Le Bonheur Children's are weaning Helen off the diet. She experienced her last documented seizure in October 2009 and today is meeting some developmental milestones.

"We treat it like medicine," said Helen's mom, Annie. "The food we give her is her medicine."

THE DIET

Versions of the ketogenic diet have been practiced since Biblical times as a way to treat seizures. Today, fats like heavy cream, butter and vegetable oils are mainstays of the diet, which allows for small amounts of lean protein and fruits and vegetables. Portions are small and all foods must be carefully prepared and weighed.

Brain chemistry is affected by the metabolic change the diet produces. Some scientists attribute the anti-seizure effect to the ketones the diet produces. The body can use these ketones as a source of energy instead of glucose, which our body normally burns for energy.

Traditionally, the diet has been used for children with myoclonic, atonic and tonic-clonic seizures.

At Le Bonheur Children's, candidates for the ketogenic diet are screened by neurologists and then referred to Ketogenic Dietitian Jennifer Jerles, MS, RD, LDN. Jerles is dedicated to treating children on the diet and performs the diet initiation, sees children in the hospital and at follow-up visits with the neurologist, and answers parent e-mails and phone calls between visits.

"She is part of a multi-disciplinary team that includes a neurologist with expertise in the ketogenic diet, social workers and nurses," said James Wheless, MD, director of the Neuroscience Institute at Le Bonheur Children's.

At Le Bonheur, patients stay on the diet an average of two years, depending on how they respond, Jerles said. If they remain seizure free, the child's neurology team will wean them off slowly, slightly adjusting fat-to-carbohydrate ratios until they are off the diet.

"The ketogenic diet tends to give families a sense of hope," Jerles said. "So many families come to us seeking out this treatment. Because the diet gives them another non-medication option, families are willing to try it."

HARD WORK

In an average week, the Webers spend their Sundays carefully preparing Helen's meals for the next seven days. Everything must be precisely mixed and measured. A typical meal might be turkey and butter, or whipped cream and fruit, says mom, Annie.

They are in regular contact with Jerles, who helped develop a meal plan and shares recipes with the couple. Throughout the week, caregivers trained in feeding Helen come to her home when the Webers are at work.

"Jennifer has been really great by following up on e-mail, and she's been very accessible when I have questions," Annie said.

Helen also receives a formula mixture through her feeding tube. Because Helen is still young, the Webers have been able to use the feeding tube to replace any nutrition she doesn't receive from her meals, Annie said.

When the Webers first decided to try the ketogenic diet a year ago, they traveled from their Oxford, Miss., home to Le Bonheur, where they began the diet. At first, it was rough, Annie admits. Once Helen was able to maintain the diet, doctors began weaning her off her medications. She underwent EEG testing in the hospital's Epilepsy Monitoring Unit during this time, until she was stabilized to go home.

After just a couple of months on the diet, Helen was seizure free. A year later, the Webers say their team effort has paid off.

"There's nothing simple about this diet. We didn't go into it lightly, we knew it was a big change," Annie said. "It's a lot of work, but it does work. My advice to other parents would be: if you think it might help, you should try it."

Tic Clinic Incorporates Behavior Modification into Care

Robin Morgan, MD

A Le Bonheur neurologist is working to incorporate comprehensive, behavioral intervention into the treatment of children with tics and Tourette's Syndrome.

The goal, says Robin Morgan, MD, is to minimize the amount of medications children with these diagnoses receive and train them to control the tics with simple behavioral techniques. Morgan currently sees about 200 children in her clinics in Memphis and Northern Mississippi who suffer from tics and Tourette's.

In her clinic, Morgan takes on the role of physician and educator, helping patients and families better understand Tourette's Syndrome and how it may affect various aspects of their lives. Morgan also works with classroom teachers to help ensure the children she sees are able to integrate well at school.

She sees children with simple motor or vocal tics and those with Tourette's Syndrome. Tourette's is a clinical diagnosis based on the presence of multiple motor and at least one vocal tic present for more than one year. Many children with Tourette's and tic disorders also have symptoms of Attention Deficit Disorder/Attention Hyperactivity Disorder and Obsessive Compulsive Disorder.

"Our goal is to make the correct diagnosis, educate families and then prescribe medications if they are warranted," Morgan said. She cites a recent national study on Tourette's that compared children who were trained to control their tics using habit-reversal techniques with those who took medication to control the tics. The results for each group were the same, Morgan said.

In her clinic, Morgan says parents and children with tics are often relieved to know and understand the diagnosis and to be provided with tools to manage the symptoms. When warranted, she is also able to treat and prescribe medication for the co-morbid conditions of Tourette's, often with the help of psychiatry – like anxiety, depression, Obsessive Compulsive Disorder and Attention Deficit Hyperactivity Disorder. She is hoping to establish a multidisciplinary clinic with input and treatment provided by pediatric neurology, psychiatry and behavioral psychology.

SAVE THE DATE: *Neurology Update*

Save the date for the 5th Annual Greater Mid-South Pediatric Neurology Update on May 6-7, 2011, at the Westin Beale Street Hotel in downtown Memphis.

The seminar is designed to encompass state-of-the-art practices and trends in treating children with neurologic disorder disorders. Seminar faculty will provide insight into common situations that subspecialists in pediatric neurology face, using case-based learned and didactic lectures with question and answer time.

To learn more or to register online, call 901-516-8933 or visit www.lebonheur.org/cme.

Le Bonheur Children's Hospital
Methodist Healthcare Family

Craniofacial Team article continued from page 1

allowing the infant to grow up with a normal appearance.

Most patients are able to leave the hospital after three days following this procedure.

For the LaFountaines, knowing Boop and Wallace were in charge of their child's care made all the difference in the world.

"We were impressed with their performance history," said dad, Christopher, "and they really seemed head and shoulders more knowledgeable in this area."

Le Bonheur Children's Hospital
Methodist Healthcare Family

Brain Waves is a quarterly publication of the Neuroscience Institute at Le Bonheur Children's Medical Center. The institute is a nationally recognized center for evaluation and treatment of nervous system disorders in children and adolescents, ranging from birth defects and learning and behavioral disorders to brain tumors, epilepsy and traumatic injuries.

50 N. Dunlap Street
Memphis, Tennessee 38103

Non-Profit Org.
US POSTAGE
PAID
Memphis, TN
Permit No. 3093

Study Tests Bed Alarm in Detecting Seizures

Neurologists at Le Bonheur Children's are studying the efficacy of nocturnal seizure alarms in detecting seizure activity in a pediatric population.

The study will determine if a specific bed alarm that fits under a child's mattress can accurately detect various seizure types. Researchers also hope to find what seizure types are best detected with the system, the rate of false alarms and if parameters for detection can be established for all ages and body weights of childhood.

"If we could say this is a device that works well for children, we could ease the minds of parents of children with known or suspected night seizures," said Kate Van Poppel, MD, a neurophysiology fellow in Le Bonheur Children's Neuroscience Institute. "These parents now use baby alarms, apnea monitors and pulse oximeters to monitor their children at night, and these methods aren't perfect by any means."

Van Poppel and her colleague, neurophysiology fellow Stephen Fulton, MD, are conducting the research and hope to enroll 100 patients. The study began in July in Le Bonheur's Epilepsy Monitoring Unit where the seizure alarm is placed in the bed according to manufacturer standards. Patients also have standard monitoring in place, including video electroencephalography, cardiopulmonary monitoring and nursing staff monitoring. The established monitoring can be compared to the new alarm system to determine its effectiveness.

The alarm device under study detects ongoing seizure activity by monitoring for prolonged rhythmic movements, sound or change in breathing movements. The system connects via radio signal to a portable pager for caregivers to keep with them. The alarm is activated after five seconds of continuous movement or sounds, and caregivers are alerted through pagers.

Current methods of monitoring – like apnea monitors and pulse oximeters – have frequent false alarms that can lead to even more anxiety for the caregiver.