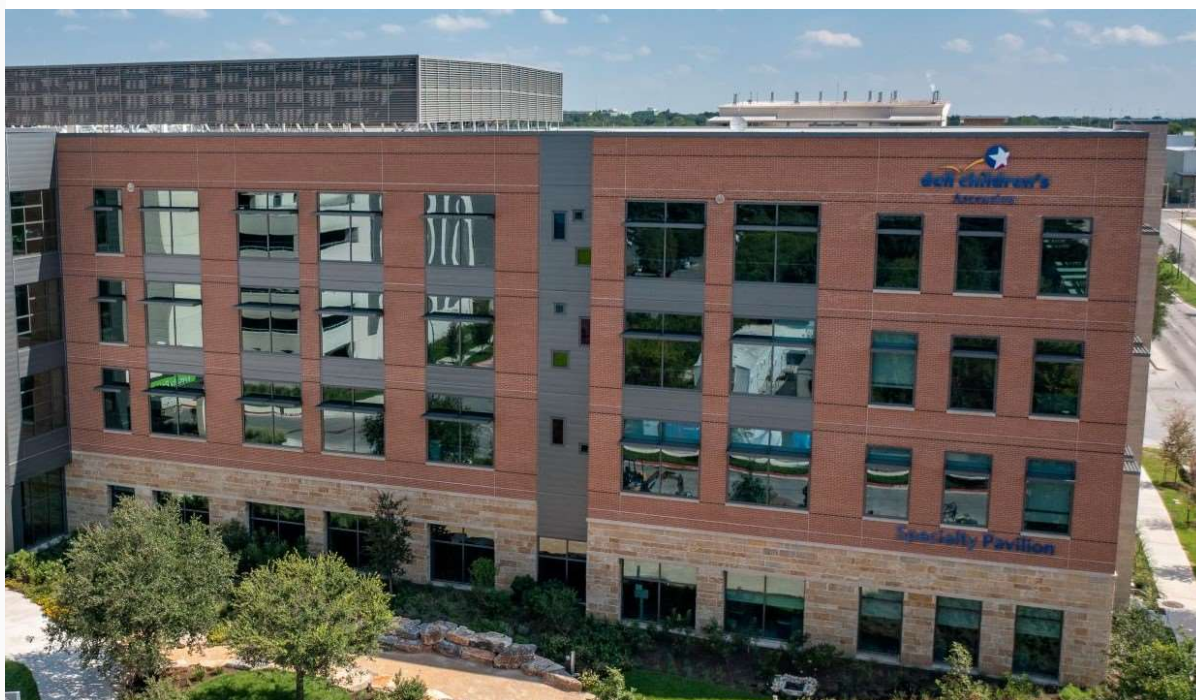


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# The Neurotransmitter

UT Health Austin Pediatric Neurosciences at Dell Children's

Promoting Discovery and Innovation in the Pediatric Neurosciences



Dear colleagues:

Welcome to UT Health Austin Pediatric Neurosciences at Dell Children's. We believe this first *Neurotransmitter* newsletter will provide a sense of the excitement surrounding our rapidly growing new program. We are committed to offering outstanding educational opportunities, promoting research and scholarly activity, and delivering exceptional multidisciplinary clinical care for all children with neurological disorders. We are always happy to hear from you!

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## **DRAMATIC GROWTH OF THE PEDIATRIC NEUROSCIENCES PROGRAM CONTINUES**

The **UT Health Austin Pediatric Neurosciences Program at Dell Children's** has grown dramatically since it was founded in 2019. The multidisciplinary program currently features 15 child neurologists, three pediatric neurosurgeons, a pediatric physical medicine and rehabilitation specialist, six pediatric neuropsychologists, and a pediatric neuro-ophthalmologist. We recently welcomed several new Dell Medical School faculty members to the program:



**Duriel Hardy, MD**, assistant professor of Neurology at Dell Medical School, is a pediatric neuroimmunology specialist. He is a graduate of Duke University School of Medicine and completed a child neurology residency and a fellowship in multiple sclerosis and neuroimmunology disorders at the Children's Hospital of Philadelphia and the Hospital of the University of Pennsylvania.



**Glendaliz Bosques, MD**, associate professor of Neurology, is chief of Pediatric Rehabilitation Medicine at Dell Children's Medical Center. After receiving her medical degree from the University of Puerto Rico, Bosques completed residency training in PM&R in Houston at the combined Baylor/UT Health Science Center program. She then did a fellowship in pediatric PM&R at Cincinnati Children's Hospital. Before joining the Dell Medical School faculty, Bosques served as chief of Pediatric Rehabilitation Medicine at Memorial Hermann and the Shriners Hospital for Children in Houston. She brings a wealth of experience and a strong vision for our pediatric PM&R program.



**Sarah Weatherspoon, MD**, associate professor of Neurology at Dell Medical School, is an epilepsy specialist who has a particular interest in infantile spasms and the neurological disorders of neonates. She is a graduate of the University of Texas Southwestern Medical School in Dallas and completed residency training in child neurology and a fellowship in pediatric epilepsy at Cincinnati Children's Hospital. Before joining the Dell Medical School faculty, Weatherspoon was on the faculty of the University of Tennessee Health Science Center in Memphis.



**Sara Pavitt, MD**, assistant professor of Neurology at Dell Medical School and chief of the Pediatric Headache Center at Dell Children's, specializes in the diagnosis and management of headache in children and adolescents. Pavitt received her medical degree from the Rosalind Franklin University of Medicine and Science in Chicago, then completed a child neurology residency at Stanford University. Prior to joining the faculty of Dell Medical School, she completed a pediatric headache fellowship at the University of California, San Francisco.



**Vandana Vedanarayanan, MD**, assistant professor of Neurology at Dell Medical School, is a general child neurologist and educator. She completed medical school and child neurology residency training at the University of Mississippi in Jackson, Mississippi, and served on the medical school's faculty prior to joining the faculty of Dell Medical School.



**Stephen Deputy, MD**, professor of Neurology at Dell Medical School, focuses on Tourette syndrome, tics, and other movement disorders. He received his medical degree from Northwestern University Feinberg School of Medicine in Chicago and completed residency training in pediatric neurology at the University of California, San Francisco. Deputy joins the Dell Medical School faculty after several years at Louisiana State University Health Sciences Center in New Orleans, where he won numerous student and resident teaching awards.

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## **PANDEMIC ADAPTION OF NEUROPSYCHOLOGICAL PRESURGICAL TESTING**

The COVID-19 pandemic altered virtually every facet of health care. Nonemergency procedures were postponed to preserve hospital bed capacity, routine health care was often deferred, cumbersome patient and provider screening procedures were implemented, and telemedicine was embraced to reduce the exposure risk for both patients and health care workers.

The presurgical neuropsychological evaluation of children with drug-resistant epilepsy is particularly challenging under these conditions. The standard test instruments have only been validated for in-person use, and we cannot assume that remotely administered tests are equally valid. Environmental distractions are more difficult to control when the patient is at home. Cooperation with testing is improved by establishing rapport with the patient and family, but this is more difficult with remote testing. The remote examiner could also miss subtle clues about the child's effort level or understanding the task at hand.

**Nancy Nussbaum, PhD**, and her colleagues at Dell Medical School explored these pandemic-related challenges as they related to pediatric patients with drug-resistant epilepsy, analyzing evidence-based methods of virtual evaluation to develop a systematic decision-making process for remote neuropsychology. In this article they provide several scenarios to guide the use of virtual neuropsychological assessment and discuss the potential pitfalls of remote testing. The need for remote test validation remains, of course, but in the meantime, the models presented here may be useful for other multidisciplinary surgical teams who need to utilize remote neuropsychological testing.

Nussbaum NL, Young SR, DeLeon RC, Engelmann ML, Schraegle WA. [The future is now: pediatric neuropsychological presurgical epilepsy evaluation in the age of COVID-19. \*Epileptic Disord\* 2021;23:274-280.](#)

## **PEARLS & POTHOLES**

This eight-year-old boy's family requested a second opinion several months after his diagnosis of Friedreich ataxia. The diagnosis was based on his physical findings and electrodiagnostic evidence of a sensorimotor axonal neuropathy. Gene testing had failed to confirm Friedreich ataxia. In the months following his diagnosis, his ataxia rapidly worsened, he developed hypersomnolence, and his once-stellar school performance deteriorated. His medical history was notable for chronic proteinuria and long-standing attention deficit disorder.

Abnormal examination findings included mild dysarthria, an ataxic gait, a positive Romberg sign, bilateral foot drop, and bilateral Babinski responses. Vibratory and position sensation were diminished distally in the legs, and pinprick and temperature sensations were diminished distally in all extremities.

What other diagnoses should be considered?

*See below for additional discussion.*

## **LASER INTERSTITIAL THERMAL THERAPY ADAPTED FOR INFANTS**

**Dave Clarke, MD, Elizabeth Tyler-Kabara, MD, PhD, and Winson Ho, MD**, of the Pediatric Neurosciences Program recently performed laser interstitial thermal therapy on a four-month-old infant. This is likely the youngest patient to undergo the procedure.

Although laser interstitial thermal therapy is increasingly utilized in the surgical treatment of epilepsy, it is seldom employed in individuals less than two years old. Placement and stabilization of the laser fibers rely on stereotactic navigation techniques that assume maturity of the patient's skull configuration. However, laser interstitial therapy is a particularly intriguing option for individuals who may have multiple epileptogenic zones that could require multiple procedures over time, such as individuals with tuberous sclerosis complex (TSC).

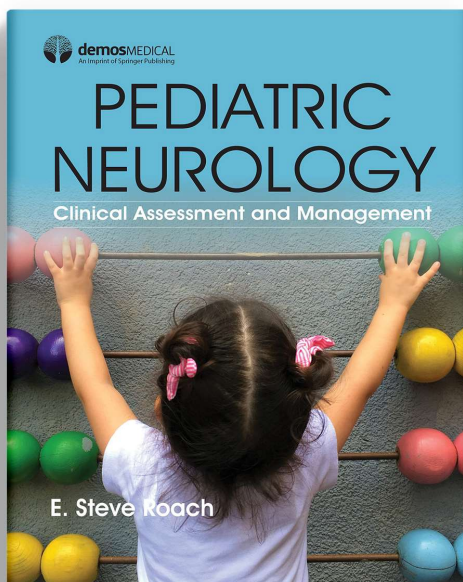
The team adapted a Navigus biopsy skull mount and used electromagnetic navigation to position the laser fiber in an infant. The four-month-old child with TSC had experienced numerous clinical and electrographic seizures per day. Noninvasive testing (MRI, magnetoencephalography, scalp EEG, and SPECT) localized the seizure onset to a left frontal premotor cortical tuber, but several other tubers were present. The child underwent successful laser ablation of the epileptogenic tuber and remained seizure-free several months after surgery.

This new technique shows the feasibility of laser surgery for very young patients and should be particularly useful for individuals who might require multiple procedures.

Lee JJ, Clarke D, Hoverson E, Tyler-Kabara EC, Ho WS. [MRI-guided laser interstitial thermal therapy using the Visualase system and Navigus frameless stereotaxy in an infant: technical case report.](#) *J Neurosurg Pediatr* 2021;1-4.

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## NEW PEDIATRIC NEUROLOGY TEXTBOOK RELEASED



Edited by **E. Steve Roach, MD**, of Dell Medical School, the recently published *Pediatric Neurology: Clinical Assessment and Management* is an introductory text for trainees and other individuals who need basic information that is presented in a clear, straightforward fashion.

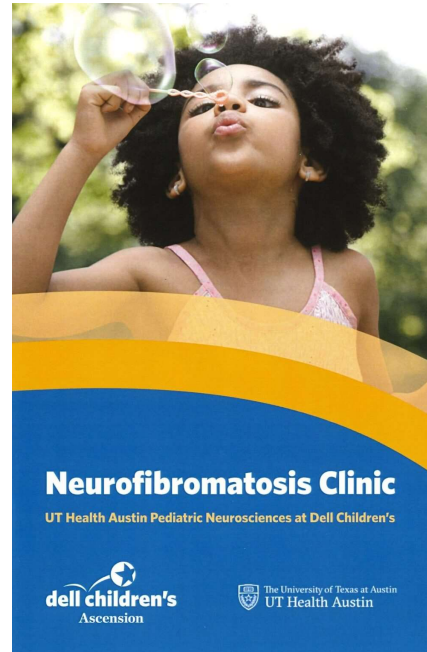
The book includes chapters contributed by numerous leading child neurologists. Beginning with an overview of the clinical assessment of infants and children, the book includes sections on the major types of neurological disease of children and features up-to-date disease guidelines and evidence-based treatment recommendations.

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## NEUROFIBROMATOSIS CLINIC JOINS THE CHILDREN'S TUMOR FOUNDATION NF CLINIC NETWORK

Our Neurofibromatosis Clinic has been accepted as a member of the **NF Clinic Network of the Children's Tumor Foundation** — a major accomplishment for a new clinical program. The NF Clinic Network recognizes clinics that provide comprehensive medical care to individuals with NF, foster patient education, and encourage participation in clinical research trials and the NF registry.

Our clinic is co-directed by pediatric neurologist **Manikum Moodley, MD, FRCP**, who joined us in 2019 after serving for several years as co-director of the Cleveland Clinic Neurofibromatosis Center, and pediatric neuro-oncologist Virginia Harrod, MD, PhD. Moodley and Harrod have extensive experience with the diagnosis and management of neurofibromatosis. Our clinic coordinator is **Karla Robles Lopez, MD, PhD**, from the Pediatric Neurosciences Program staff. This program is one of several multidisciplinary clinics developed by UT Health Austin Pediatric Neurosciences at Dell Children's.



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## OUR COLLEAGUES IN THE NEWS

### **Audrey Brumback Named to CNS Leadership, Diversity, Equity, and Inclusion Task Force**



**Audrey Brumback, MD, PhD**, was recently appointed to the newly formed Child Neurology Society Leadership, Diversity, Equity, and Inclusion (LDEI) Task Force. Brumback also serves as an elected member of the CNS board of directors. The LDEI committee has already designed several strategic initiatives to begin their work of making the field of child neurology diverse, equitable, and inclusive. "We're just getting started. The goal is to bring about lasting change in everything from patient care to medical education and research," Brumback said. Thank you, Dr. Brumback, for giving your time to this important work and to the Child Neurology Society.

Brumback AC, et al. **Introducing the Child Neurology Society Leadership, Diversity, Equity, and Inclusion Task Force.** *Ann Neurol* 2021;90:537-538.

### **Dave Clarke Honored as Top 10 Physician**



**Dave F. Clarke, MD**, chief of the Comprehensive Pediatric Epilepsy Center at UT Health Austin Pediatric Neurosciences at Dell Children's, was named one of the 2021 top 10 physicians in the Austin area by the *Austin Black Business Journal and Community News*. The journal recognizes physicians who have achieved professional excellence and who have actively paved the way to success for others. Clarke and the other awardees were honored at a reception on November 20, 2021. Congratulations, Dr. Clarke!



## PEARLS & POTHOLES: EPILOGUE

Even discounting this child's unremarkable initial gene testing, his somnolence, worsening cognitive impairment, and rapid progression of clumsiness are atypical of Friedreich ataxia. A complete blood count (done to evaluate fatigue) revealed macrocytic anemia, leading to the discovery of a low serum cobalamin level.

Acquired vitamin B<sub>12</sub> deficiency in children results more often from vitamin malabsorption than from either dietary deficiency or pernicious anemia. This child, however, has selective vitamin B<sub>12</sub> malabsorption with proteinuria (Imerslund-Grasbeck syndrome), an autosomal recessive defect of the receptor that facilitates absorption of the intrinsic factor–cobalamin complex in the distal ileum. The two receptor subunits, cubilin and amnionless, are coded by the genes *CUBN* and *AMN* on chromosomes 10 and 14. A mutation of either gene can cause the syndrome.

Parenteral B<sub>12</sub> replacement corrects the hematologic abnormalities and halts the progression of the neurological manifestations, although delayed therapy does not always reverse long-standing neurological dysfunction. Oral vitamin therapy is unreliable, and treatment must be lifelong. A similar phenotype results from vitamin E deficiency, which can also result from acquired malabsorption disorders or from genetically mediated absorption defects.

### Diagnosis

Further testing confirmed compound heterozygosity for pathogenic variants in *CUBN*. With parenteral B<sub>12</sub> therapy, his hematological abnormalities resolved and his neurological dysfunction improved dramatically but did not completely resolve. Genetic disorders of cobalamin absorption are rare but highly treatable conditions, especially if diagnosed early.

### Additional Reading

Grasbeck R. Imerslund-Grasbeck syndrome (selective vitamin B<sub>12</sub> malabsorption with proteinuria). *Orphanet J Rare Dis* 2006;1:17.

Harding AE, Matthews S, Jones S, et al. Spinocerebellar degeneration associated with a selective defect of vitamin E absorption. *N Engl J Med* 1985;313:32-35.

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UT Health Austin Pediatric Neurosciences at Dell Children's is a clinical partnership between Dell Children's Medical Center and UT Health Austin, the clinical practice of the Dell Medical School at The University of Texas at Austin.

For additional program information:

[UT Health Austin Pediatric Neurosciences at Dell Children's](#)  
[Comprehensive Pediatric Epilepsy Program](#)  
[Pediatric Neurosciences Blog](#)

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