Georgia Chapter

# The GEORGIA Pediatrician

A Publication of the Georgia Chapter of the American Academy of Pediatrics

# President's Letter Harnessing Your Inner Superpower!



Terri McFadden, MD

I'll readily admit it. I am a pediatrician of "a certain age" so I'm not always up on all the latest trends. I am often the recipient of the surreptitious eye roll when I haven't learned about the latest cultural phenomenon. So, you will not be surprised to learn that when first confronted with the following question from one of my teen patients, "Which superhero franchise is

better, Marvel or DC?" I was pretty clueless. Over the years my younger daughter, a student of all things Superhero, has helped me understand the world of Superheroes and the big budget movies that accompany them, although I confess that I still don't completely understand all that happened in Avengers Endgame! Forced to watch more than my share of these films and study the intricacies of each character so as not to appear completely unintelligent, I understand enough about them to hold a very superficial conversation. For example, I understand that Spiderman can jump really far and stick to buildings and other large objects. Even Boomers and Generation X'ers know that Superman is "faster than a speeding bullet, and able to leap tall buildings with a single bound!" But more recently I have learned about Aquaman, Thor, and Ironman. And of course, there is T'Challa, the Black Panther, who moves with confidence around the nation of Wakanda and defeats all manner of evil villains bent on destroying the world. These characters and the many others that you may or may not be acquainted with, all have one thing in common. It is their superpower. Superheroes always use their powers for good and not evil. Although mere mortals have no ability to outrun speeding trains and balance wayward satellites in the palms of our hands, we do have superpowers. You aren't convinced? Well who else can manage to take one look a sick toddler's rash and make a diagnosis? Who else can have a lively conversation with a teen and actually get to the bottom of what's causing his panic attacks? Who can rescue a preschooler in the midst of a severe asthma attack? And who else can wrangle an EMR into submission, albeit while uttering a few choice words. The point is that we all have our inner superpowers. As we enter 2020 and the ever-growing list of issues that we are forced to address,

What are these superpowers, you might ask? Let me offer a few of them for your consideration...

it is so important that we harness our inner superpowers. Our patients, families and communities are counting on us to use them. What are these superpowers, you might ask? Let me offer a few of them for your consideration. I will start with my personal favorite.

- 1) Humor/Laughter: On any given day, there are many challenges that each and every one of us face. Some are serious and some are just annoying. While we don't always have control over the challenges that come our way, we do have control over our responses. I have learned to laugh at the situation and at myself. Humor is often cathartic. It is said that "he who can laugh at himself, shall never cease to be amused!"
- 2) Compassion: I know that almost all of us use this skill on a daily basis. Sometimes the sleep deprived parents who have listened to their child cough all night long, just need to hear that we acknowledge their fear, fatigue, and sense of helplessness. They just need just a little bit of affirmation as they tackle one of life's most difficult jobs. Colleagues and staff can use our compassion as well!
- 3) Calm: During the most chaotic days, having the ability to calm and center ourselves allows us to reset and get through clinically and emotionally trying situations.

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Continued from previous page.

- 4) Energy: This one can be especially challenging for most of us, but the folks who seem to have mastered the energy equation have made a conscious effort to do so. They are intentional about restoration and rejuvenation. This may result from sufficient amounts of unplugging, mindfulness, meditation, prayer, exercise or quiet time.
- 5) Empathy: While we may not be able to completely identify with the home life, personal situation, or clinical struggle of each of our patients and their families, having the ability to listen without judgement is a learned skill that must be practiced and delivered with intention.

As we begin a new decade, we will need to harness every one of our superpowers to battle for the health and safety of our children and their families.

- 6) Courage: I am inspired by quotes about courage from Nelson Mandela and Mark Twain. Per Mandela, "I learned that courage was not the absence of fear, but the triumph over." Twain said, "Courage is resistance to fear, mastery of fear—not absence of fear."
- 7) **Kindness:** This one comes naturally to most pediatricians. Most of us who work with children and families are kind people. But fatigue, stress, and lack of appreciation can leave us depleted and erode our ability to be kind.

I won't belabor the point, but I will toss out a few additional examples of inner superpowers for your consideration, such as, generosity, perseverance, gratitude, patience, spirituality, resourcefulness, wit, enthusiasm, and boldness to name a few. You might call these concepts character traits, and I won't argue, but I like to think of these traits as superpowers because I imagine placing my hands on my hips, leaning back, looking upwards, taking a deep breath, and saying to myself (as my cape floats in the breeze) "Let's do this!" In other words, using your inner superpowers is an active and intentional process. It is a decision that we make each day in our practices, clinics and hospitals. Whatever you elect to call them, these traits keep us moored and buoyed. They allow us to continue to do the noblest, most important work that there is...giving voice to our most valuable resource—our children. As we begin a new decade, we will need to harness every one of our superpowers to battle for the health and safety of our children and their families. Put on your capes and let's do this!

THE GEORGIA PEDIATRICIAN • WINTER 2020 ISSUE

Seri WfaddenHD

Terri McFadden, MD, FAAP



# How Do You Handle Transition and Change?

In the seven years I've been with

my employer, I have practiced in

three locations, and I've worked

and 1 nurse practitioner. I have

changes. As it goes, my pediatric

practice is growing, and watching

encountered several staffing

children grow is amazing!

with two pediatric partners



After residency, I began my career in a community health center. It was very busy and rewarding work, but, with a growing family that included three young children, my husband and I decided to relocate to be closer to our extended family. Seven years ago, we moved to our

current city, and it seems that the only thing that has remained constant is our home address. My husband, an educator, worked at four different elementary schools prior to settling into his new job as a college professor. In the seven years I've been with my employer, I have practiced in three locations, and I've worked with two pediatric partners and 1 nurse practitioner. I

have encountered several staffing changes. As it goes, my pediatric practice is growing, and watching children grow is amazing!

My husband and I have a 12-year marriage that has weathered the test of time, strengthened by great marriage counseling and awesome Winshape Retreats. In a time when things are ever changing, our shared faith and commitment have grounded us. Our children have grown from babies to toddlers to school-aged, and our eldest will enter middle school this fall.

Perhaps what I love most about change is that each step brings joy. I remember the excitement and pure bliss of being a newlywed and signing my name Mrs. Sylvia Washington. I remember the simple joy of breastfeeding when my kids were babies. At the end of a long on-call shift, I would take a shower, grab my Boppy, and nurse them to sleep. I remember my toddlers with their sticky hands, sippy cups, and snack cups. Their pre-school programs were precious; I especially loved watching my kids play soccer and piano.

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The thing I hate the most about change is the sorrow and grief that comes with saying good-bye. I still remember the first patient I lost, an infant in the resuscitation room, born at 20 weeks. I can picture her father's cries for help, begging for our team to "do something!" I can still hear

> that mother's scream. I remember the lessons learned from my pediatric partner who died from cancer. I made a quilt for my other pediatric partner who cared for my children for six years; she married and moved out of state. And after the privilege of guiding a nurse practitioner through her first job after graduation, she too is moving on in her life.

In my life, when there are times of transition and change, I remember the serenity prayer: God, grant me the serenity to accept the things I cannot change, courage to change the things I can, and wisdom to know the difference.

So, remember that no matter the transition or change, you are not alone! As the Chair of Early Career Physicians, I understand that change is part of life, both bitter and sweet. My goal is for every young pediatrician in Georgia to feel connected and heard. Please let me know how I can best serve you. Sending a little love your way! Hug your family for me.

Sylvia Washington, MD, FAAP Chair, Early Career Physician Section, Georgia AAP

Floyd Primary Care, Rome

# GROWING OUR TEAM AND LEADING THE WAY IN PEDIATRICS

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### Neurology



Sonam Bhalla, MD Pediatric Neurologist

**Sonam Bhalla, MD,** joined Children's by way of New York, where she completed a fellowship in pediatric epilepsy at Columbia University Medical Center. Prior to her fellowship training, Dr. Bhalla earned a medical degree at Mahatma Gandhi Mission's Medical College in India, followed by her pediatrics residency training and chief resident year at the University of Louisville. Dr. Bhalla has experience and interests in the field of pediatric neurology.



Jasmine Forte, MD Pediatric Neurologist

Jasmine Forte, MD, joined Children's after serving as Chief Pediatric Neurology Resident at Emory University School of Medicine. Prior to her residency training, Dr. Forte earned a medical degree from Meharry Medical College in Nashville, Tenn., graduating with honors. Dr. Forte has experience and interests in the field of pediatric neurology including but not limited to headache medicine and concussion.

### Psychology ———



Elyssa Gerst, PhD Pediatric Neuropsychologist

**Elyssa Gerst, PhD,** joined Children's by way of Washington DC, where she completed her postdoctoral fellowship in clinical neuropsychology at Children's National Health System. Prior to her fellowship training, Dr. Gerst completed her PhD at the University of Houston and her predoctoral internship in clinical psychology at Children's Hospital Colorado in Denver. Dr. Gerst has interests in cognitive development in children with complex medical histories, the impact of congenital or acquired neurological disorders on neuropsychological functioning, and supporting optimal outcomes in children and adolescents.



Laura Wright, PhD Pediatric Psychologist

Laura Wright, PhD, earned her doctoral degree in clinical psychology from Georgia State University and completed her predoctoral internship at Cincinnati Children's Hospital Medical Center. Given her interest in pediatric pain, Dr. Wright completed her postdoctoral fellowship at Boston Children's Hospital/ Harvard Medical School where she worked in the Pain Treatment Service, Mayo Family Pediatric Pain Rehabilitation Center and Pediatric Headache Program. Dr. Wright's primary research and clinical interests in the field of pediatric psychology include pain assessment and treatment, pediatric chronic pain, and the promotion of resilience and adaptation among youth who experience pain.



### Resident Reflection



Katherine McGlamry, MD

I sat on the floor with John in my lap. He had been dropped off the day before by his dad, a single father who had traveled many hours by public bus to bring John to the Hogar de San Francisco de Asisi, a home for ill and destitute

children located about 30 miles outside of Lima, Peru. John had not stopped crying since his arrival and was only content when sitting in a volunteer's lap. At four years old, John could not walk or talk. He was very bright and could communicate rather effectively with voice inflections and hand gestures and could crawl as fast as lightning, but he was significantly delayed and had never seen a doctor or specialist. His father could not afford medical care for John but wanted a better life for his son, so he made the difficult decision to send him to live the at Hogar, where the father would not be able to visit

his son often.

I have more empathy and understanding of the daily lives of children with complex chronic medical conditions and the struggles they and their families go through to access care.



The Hogar de San Francisco de Asisi is run by Dr. Anthony Lazzara, a former Emory University neonatologist who left his position in 1983 and moved to Peru to help children in need. Children of all ages with a variety of illnesses move into the home and receive all medical care, food, lodging and education free of charge. They stay at the Hogar as long as they need to receive care – some stay a few weeks, some a few years, some have been at the Hogar almost their entire lives. The children are taken into Lima for subspecialty appointments, surgeries and therapy sessions and are cared for 24 hours a day, 7 days a week by Dr. Lazzara and his nursing staff.

As part of the Emory Pediatrics Global Health Track, I was fortunate enough to spend a month volunteering at the Hogar de San Francisco de Asisi. I carried children on the public

buses to and from appointments in Lima, helped with chores around the house and entertained babies. I witnessed the day to day struggles that kids with chronic illnesses face in Peru. I experienced how difficult it is to get a medically complex

child to appointments on time when you are taking public transportation for hours to do so.

I was exhausted after my time at the Hogar, but it was all worth it to get to know these special children and see their progress. The Hogar is a three-story home with the bedrooms on the second floor, so I developed much stronger arm muscles carrying the wheelchair bound children up and down the stairs. I have more empathy and understanding of the daily lives of children with complex chronic medical conditions and the struggles they and their families go through to access care. Many of

my patients in Atlanta face similar obstacles to accessing healthcare, including access to reliable transportation and high costs of care. After my time in Peru, I think more about the lives of my patients outside of the office visit or hospital stay and brainstorm what I can do to help alleviate some of their burden.

John did eventually stop crying and ultimately thrived at the Hogar. He was diagnosed with Spastic Cerebral Palsy and Dr. Lazarra worked tirelessly to get him plugged in with Neurology and Orthopedic Surgery as well as physical, occupational and speech therapy. By the time I left for Atlanta, he was walking short distances independently and had picked up a few key words to go along with his hand gestures.

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# Nutrition Update Winter 2020



y Hochman, MD

Several important nutrition advances and articles have been published recently which may be of interest to Georgia pediatricians.

 At this year's Georgia AAP Nutrition Symposium, the keynote speaker, William Balistreri, highlighted "The Paramount Health Challenge for Humans in the 21st Century" and focused attention on a recent study (GBD 2017 Collaborators. Lancet 2019; 393: 1958) which attributed 20% of deaths worldwide as due to poor diet.

A link to a summary and slides from this lecture:

• gutsandgrowth.com/2019/10/17/the-paramount-health-challenge-for-humans-in-the-21st-century

Here's a link to the full Lancet Article pdf:

• thelancet.com/pdfs/journals/lancet/PIIS0140-6736(19)30041-8.pdf

### Effects of unhealthy diets - Global Burden of Diseases (GBD) study GBD 2017 Collaborators, Lancet 393:1958, 2019

- Evaluated consumption of foods & nutrients in 195 countries; quantified impact of suboptimal intake
- ~20% of deaths (~11M) worldwide assoc with poor diets; most deaths due to CV disease, cancer, Type 2DM

Another good commentary on the health effects of our diet from the NY Times: "Our Food is Killing Too Many of Us" (August 26, 2019)

 nytimes.com/2019/08/26/opinion/food-nutrition-healthcare.html?smid=nytcore-ios-share

The Georgia AAP Nutrition Symposium also featured another excellent lecture from William Balistreri on food safety. A link to a summary and slides from this lecture:

- gutsandgrowth.com/2019/10/18/food-safety-lecture-it-is-still-a-jungle-out-there
- 2. In May 17, 2019, the FDA approved Glucagon-Like Peptide-2 (GLP-2) for use in pediatric patients with short bowel syndrome. GLP-2 mimics naturally occurring glucagon-like peptide-2 (GLP-2), which helps the remaining intestine absorb more nutrients. GLP-2 was approved based in part on a recent study (SA Kocoshis et al JPEN 2019; doi: 10.1002/jpen.1690). At the end of the 24-week study, 69% of patients (18/26) who took GLP-2 (0.05 mg/kg each day) reduced parenteral support (PS) volume by 20% or more; in those receiving 0.05 mg/kg/day, PS dropped by a

mean volume of 42% (mL/kg/day) from baseline (-23 mL/kg/day from baseline). At week 24, 38% of patients (10/26) were able to reduce PS infusion by at least 1 day per week. Patients reduced their PS infusion time by 3 hours per day on average compared to baseline. In addition, during this study 3 out of 26 (12%) children who received GLP-2 (0.05 mg/kg/day) completely weaned off PS.

#### My take:

There are many limitations before this medication is adopted:

- a. It is a very expensive medication (cost in adults ~\$295,000/year)
- b. The effects may wear off when the medication is stopped
- c. More time is needed to confirm its safety profile.
   Current recommendations would impose additional monitoring (e.g. Colonoscopy at 1 yr and then every 5 yrs) for patients receiving GLP-2
- 3. A few years back, the general advice was to avoid prolonged sitting at work as important for good health. A more recent study (JM Garcia et al. Journal of the American Heart Association; 2019; doi.org/10.1161/JAHA.118.010406) challenges that advice.

#### **Key findings:**

- "After adjusting for health and lifestyle factors, the researchers found that "often or always" sitting at work was not associated with a heightened risk of death and heart disease"
- "But those watching four or more hours of television per day had a 50% higher risk of heart issues and death, compared to those watching two hours or less per day."
- "The health risk of watching lots of TV vanished when people spent 150 minutes or more per week doing moderate-to-vigorous exercise— like brisk walking, running, swimming and cycling."

### My take:

This study suggests that sitting a lot at work is mainly a problem only for those who sit a lot when they leave work.

### Jay Hochman, MD

Past Chair, Committee on Nutrition Georgia AAP Blog site: gutsandgrowth.wordpress.com



# Can We Handle the Truth?



Robert Wiskind, MD, FAAP

A Few Good Men (1992) is a courtroom drama centered on the murder trial of two Marines charged with murdering a colleague. The Marines claim that their commanding officer, Colonel Jessup, ordered the severe form of discipline ("Code Red") that led to the death. In the absence of direct evidence, the Marines' attorney attempts to badger Colonel Jessup into admitting he ordered the Code Red, claiming that the court is entitled to the answer and the truth. Jessup's memorable reply: "You can't handle the truth!!"

Despite the current political climate and the Internet blurring the lines between fact and fiction, the truth is still out there (to misquote The X-Files). As a nation, the state of Georgia and pediatricians, we do ourselves and the children we care for a disservice when we fail to

acknowledge these truths.

President Trump claims we have no option but to mistreat children who entered the U.S. without documentation; the truth is that all children's health and safety should be a priority

We must be able to demonstrate the value of the care we provide, measured by clinical outcomes that maintain the health of a population of children.

and that our society will be judged by how we care for those less fortunate who can't care for themselves. The state of Georgia has not fully expanded Medicaid under the ACA, claiming that we can't afford to make sure that all residents have affordable health insurance, while the truth is that the federal program will bring \$9 to Georgia for every \$1 that the state spends in expanding Medicaid and that uncompensated care is financially crippling hospitals and physicians. At the state and national level, leaders say they care about protecting children from gun violence, vaping and drugs, yet the truth is they lack the fortitude to stand up to the NRA, tobacco manufacturers and others who believe their rights outweigh the rights of children to be healthy and safe. As citizens and voters, we have a duty to speak the truth to politicians and insist they do what is right, not

Every pediatrician I know thinks that they practice evidencebased medicine and provide the best possible care to their patients. The truth remains that until you closely examine your practices and outcomes, you are just guessing. Quality improvement (QI) is much maligned, especially by those who only see it as a component of Maintenance Of Certification

what is easy or benefits them politically.

(MOC). The truth is that QI is the future of medicine. We must be able to demonstrate the value of the care we provide, measured by clinical outcomes that maintain the health of a population of children. We must assess how we are preforming compared to peers and benchmarks, make meaningful changes when we don't measure up and then reassess to document improvement. The Children's Care Network (the Atlanta-based Clinically Integrated Network) has shown that regular use of Asthma Action Plans (which some physicians deride as 'busy work') decreases the rates of ER visits and hospitalizations for asthma exacerbations. Recent preliminary data has also shown that routine screening for Depression increases the number of teens diagnosed with mental health problems while simultaneously lowering

their rates of ER visits and hospitalizations. The truth is that our profession needs to adapt to the demands of valuebased healthcare; if we don't, we risk becoming extinct.

The truth is that once a pediatrician has passed initial Board Certification, there is no guarantee that she/he will

maintain the skills and knowledge to care for children safely and effectively. State licensure is not a measure of competence and CME is not sufficient to document knowledge; any certification process that relies solely on licensure and CME is inadequate. MOC is necessary to demonstrate that pediatricians remain competent. The AAP and individual Pediatricians should continue to work with the American Board of Pediatrics to improve the MOC process.

After Colonel Jessup admits he ordered the Code Red, the two Marines are acquitted of murder, but found guilty of conduct unbecoming a Marine and given dishonorable discharges. One complains that they were only following orders, but the other replies that they should have stood up for their fellow Marine, stating "We're supposed to fight for people who can't fight for themselves." Acknowledging the truth about our society and ourselves will help pediatricians continue to fight for children every day.

Robert Wiskind, MD, FAAP

Past President, Georgia AAP Peachtree Park Pediatrics, Atlanta

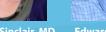
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### A Charged Topic: Button Battery Ingestions







Erica L Riedesel, N

Surveillance data from the National Capital Poison Center (NCPC) has demonstrated a dramatic rise in US emergency department visits for pediatric button battery ingestion (BBI) over the past 10 years (Fig 1).

Button batteries are found in a number of household items such as toys, greeting cards, remote controls, watches, and key fobs. Today, many of these items use larger coin-size 3-volt lithium batteries. When swallowed by children, these batteries can become lodged in the esophagus. Battery current reacts with saliva and generates hydroxide, creating a caustic (alkaline) injury to the esophageal tissue (Fig 2). Severe injury can occur in as little as two hours. The longer the battery remains in the esophagus, the more severe the injury, sometimes resulting in full thickness damage to the esophageal wall, perforation, and even erosion of the adjacent airway or adjacent blood vessels leading to potentially fatal complications.

Prompt medical attention is needed if a child is suspected of having swallowed a button battery; however, ingestions are often unwitnessed and diagnosis cannot be made by history and physical exam alone. Toddlers cannot distinguish between a coin and a button battery and many of the symptoms of BBI can mirror other illnesses. X-rays are essential and must include frontal and lateral images from the nose to the rectum. Look for the distinctive "halo rim" on frontal x-ray and "step off" edge on lateral x-ray (Fig 3).

Once a button battery ingestion is confirmed, every minute counts. National guidelines from the North American

Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) and NCPC recommend removal of button batteries in the esophagus within 2 hours. Button batteries in the stomach should also be urgently removed if the battery measures >20mm and the child is under 5 years of age or if the child is symptomatic. While gastric mucosal injury is rare, these patients can also have esophageal injury. Safe removal can be difficult and should be performed by pediatric specialists in Gastroenterology, Surgery, or ENT.

The healing process following button battery ingestion is complex, especially in patients with a prolonged battery exposure. These children remain at high risk and are often monitored closely in the hospital for 4-30 days with serial imaging studies to detect any late complications, such as esophageal stricture or perforation, tracheoesophageal fistula, or aortoesophageal fistula.

In the past year, a group of physicians at Children's Healthcare of Atlanta (CHOA) has become dedicated to studying how button battery ingestions are affecting children in Georgia. Our center reviewed all cases of BBI at CHOA from 2007-2018, a total of 44 esophageal button battery impactions and 114 gastric button battery cases. In our population, battery size >20mm and patient age < 6 years were significantly associated with esophageal impaction. Most button batteries were lodged in the upper esophagus. Children presented with a wide variety of symptoms and, in one third of cases, parents had no suspicion of button battery ingestion. Importantly, 15% of ingested button batteries

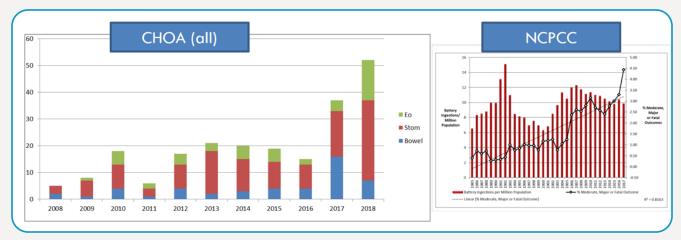


Figure 1: Local and National Rise in Button Battery Ingestions

## Button Battery Ingestions

Continued from previous page.

were misidentified as coins or tokens on initial x-ray, which resulted in delay in care. Time to battery removal was also delayed with only half of esophageal batteries removed within 2 hours of identification. Five patients (11%) developed complications including tracheoesophageal fistula and esophageal stricture.

Ensuring that children do not have access to button batteries is the best way to prevent battery-related injuries. Batteries should be stored out of children's reach and parents should check products that use button batteries to ensure the battery compartment is securely closed.

As clinicians, our community needs to work together to improve identification of button battery ingestion and timely button battery removal.



Middle Third of the Esophagus

Middle Third of the Esophagus

Figure 2:

Example of Injury due to Esophageal Button Battery Impaction

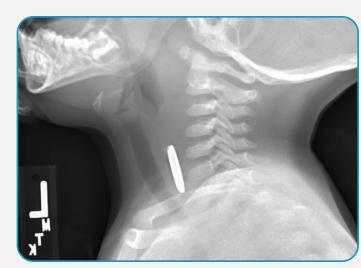


Figure 3: X-rays of Button Battery Ingestions

# When to suspect a button battery ingestion:

- Witnessed or suspected ingestion of any foreign body.
- Symptoms: Vomiting, coughing, gagging, new onset stridor, drooling, difficult or painful swallowing, refusing oral intake, blood in saliva, hematemesis, chest pain, or neck pain.

# What to do when a button battery ingestion is suspected:

- AP and lateral x-rays from the nose to rectum. Carefully assess images for characteristic "halo rim" on frontal x-ray and "step off" on lateral x-ray of a button battery.
- Call the National 24-hour National Battery Ingestion Hotline at 1-(800)-498-8666 or call your poison center at 1-(800)-222-1222.
- Call CHOA Transfer Center 404-785-7778 or 888-785-7778 to speak with a specialist in Pediatric Gastroenterology or Pediatric Surgery to arrange prompt transfer and battery removal.

### Elizabeth M Sinclair, MD

Pediatric Gastroenterology, Hepatology, and Nutrition Fellow Emory University Children's Healthcare of Atlanta

### Edward Richer, MD

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# Role of the Primary Care Physician and Medical Home in the Early Hearing Detection and Intervention Program

Congenital hearing loss occurs in

1-3 out of 1000 infants in the United

whose parents have normal hearing.

**States and is often found in babies** 



Paula J. Harmon, MD

Congenital hearing loss occurs in 1-3 out of 1000 infants in the United States and is often found in babies whose parents have normal hearing. Well-established evidence shows that early identification of hearing loss and appropriate early intervention can mitigate the potential for poor language acquisition. The American Academy of Pediatrics (AAP) endorsed the Joint Committee on Infant Hearing Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. This

statement supports universal newborn hearing screening and outlines best practices in the timing and methods for follow up hearing screening and participation in early intervention. Early detection of hearing loss facilitates language acquisition and

future opportunities for educational success.

Pediatricians and other pediatric primary care providers play a vital role in mitigating developmental delays in infants and young children who are deaf and hard of hearing. Those providing a medical home for children can ensure appropriate and timely referrals to providers, who can perform evaluations and are knowledgeable about congenital hearing loss. The Georgia Department of Public Health (DPH) provides an Early Hearing, Detection, and Intervention (EHDI) coordinator for each county to assist in follow-up services for families of infants who do not pass the initial hearing screening. You can find your local EHDI Coordinator online at: <a href="mailto:sendss.state.ga.us/sendss/!mch.coord-search.">sendss.state.ga.us/sendss/!mch.coord-search.</a>

Since its release, the EHDI program adopted the AAP Guidelines for Pediatric Medical Home Providers, available online at **pediatrics.aappublications.org/ content/120/4/898/tab-figures-data.** This algorithm maps the 1 - 3 - 6 follow-up guidelines for newborn hearing screening as shown in Figure 1. Outlined below are the key components of the physicians' role in this process:

1. For all newborns, obtain written results of newborn hearing screening from the birthing facility. Results are available via a web portal through the Georgia Department of Public Health's State Electronic Notifiable Disease Surveillance System (SendSS) or eReports. Physicians can register for SendSS and eReports by following these links <a href="mailto:sendss.state.ga.us/sendss/login.screen">sendss.state.ga.us/sendss/login.screen</a>; <a href="mailto:ereports.ga.gov/dph/eReports/weblogin.gengs">ereports.ga.gov/dph/eReports/weblogin.gengs/msgNum=0&Locale=1033</a>

2. By one month of age, ensure that all newborns receive a hearing screening. Infants who "refer" on their initial

screening, in one or both ears, must receive a follow-up hearing screen. Infants that pass their initial screening do not need additional testing unless concerns arise. If the initial hearing screen uses an automated auditory brainstem response (a-ABR)



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### Early Hearing Detection and Intervention

Continued from previous page.

screener, then the same test must be performed during the outpatient, follow-up hearing screen.

- 3. By three months of age, for infants that refer on their outpatient follow-up screen, send for a diagnostic evaluation. Complete only one outpatient screening, as repeated screenings do not provide enough information to determine appropriate follow-up recommendations. Repeated screenings delay the identification of hearing loss. After three months of age, sedation may be required for testing, which can be a deterrent for parents. EHDI coordinators can assist in locating appropriate providers, as needed.
- 4. By six months of age, support referrals to Early Intervention, Otolaryngology, Ophthalmology, and Genetics, after diagnosis of permanent hearing loss. A suspected or confirmed case of hearing loss in children from birth to age 5 is a notifiable disease and requires a report to the Georgia Department of Public Health. It is important, therefore, to notify the EHDI program upon confirmation of permanent childhood hearing loss. Send a copy of the diagnostic report to the EHDI Program or a surveillance form for reporting hearing loss for children five and under, located on the GA Chapter of the American Academy of Pediatrics website. EHDI Coordinators will work with the child's physician and family to assist in linking infants with early intervention services.
- Otitis media with effusion can lead to permanent hearing loss. Infants and children must be referred to an otolaryngologist and audiologist.
- 6. Hearing loss may develop at any age; therefore, it is important to monitor all infants for progressive or late-onset hearing loss. Referral for audiological evaluation is recommended at least once before age 30 months for infants who have risk indicators for late-onset hearing loss, such as family history of permanent childhood hearing loss, neonatal intensive care unit stay of more than five days duration, and parental concern. The attached algorithm Includes a list of risk indicators.

For more information on the Georgia EHDI Program, please visit **dph.georgia.gov/EHDI** or call 404-651-5482

For information on the American Academy of Pediatrics Program to Enhance the Health & Development of Infants and Children (PEHDIC), visit <a href="majoragen-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Early-Hearing-Detection-and-Intervention.aspx">aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Pages/Early-Hearing-Detection-and-Intervention.aspx</a> or contact Fozia Khan Eskew, Director of Child Health at the Georgia Chapter of the American Academy of Pediatrics at feskew@gaaap.org.

### Paula J. Harmon, MD

Georgia EHDI Champion Georgia Chapter - American Academy of Pediatrics



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# Forgetting to Breathe: Congenital Central Hypoventilation Syndrome





Ajay S. Kasi, M.D.

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Congenital central hypoventilation syndrome (CCHS) is a rare genetic disorder of the autonomic nervous system (ANS) and respiratory control due to a mutation in the paired-like homeobox 2B (PHOX2B) gene.¹ Affected individuals typically present in the neonatal period with apnea, cyanosis, hypoxemia, and hypercapnia that is worse during sleep but may also be present during wakefulness. Once placed

on ventilatory support, they fail to be weaned off assisted ventilation. Older infants and children can present with unexplained apneas, hypoventilation, or respiratory failure following either a respiratory infection or anesthesia.<sup>1,2</sup> Children with CCHS have

assisted ventilation, at least during sleep. Severe cases require continuous ventilatory support.

an absent or negligible ventilatory response to hypoxemia and hypercapnia during both sleep and wakefulness.<sup>3</sup> Hence, these children do not demonstrate signs of respiratory distress such as tachypnea, chest retractions, and/or nasal flaring despite profound hypoxemia and hypercapnia. This discrepancy between the clinical presentation and gas exchange abnormalities, along with the rare incidence of this disease, makes the diagnosis and management of CCHS challenging.

When the diagnosis of CCHS is suspected, PHOX2B gene mutation testing should be performed, and the patient should be evaluated for other causes of hypoventilation such as cardiopulmonary, neurologic, neuromuscular, and metabolic disorders. Evaluations include chest and brain imaging, electrocardiogram, echocardiogram, neurological and metabolic studies, and a polysomnogram. CCHS is a generalized disorder of the ANS and other clinical manifestations include Hirschsprung's disease, cardiac arrhythmias that may require cardiac pacemaker implantation, pulmonary hypertension, neural crest tumors, as well as ophthalmologic and temperature regulation abnormalities.<sup>1</sup>

CCHS patients require lifelong assisted ventilation, at least during sleep. Severe cases require continuous ventilatory support. Generally, the PHOX2B mutation genotype can aid in anticipating the severity of the phenotype. Home assisted ventilation options include positive pressure ventilation (PPV) via tracheostomy, noninvasive positive

pressure ventilation, and diaphragm pacing (DP).<sup>1,2</sup> DP generates respiration using the patient's own diaphragm by electrical stimulation of the phrenic nerves. The DP system is comprised of the following: 1) electrodes that are surgically implanted on the phrenic nerves bilaterally; 2) receivers that are subcutaneously implanted in the chest or abdomen; 3) antennae that are taped on the skin over the receivers

when pacing; and 4) a battery-operated external transmitter. (Figure 1) The transmitter generates a train of pulses, which are transmitted to the receivers by the antennae. The receivers convert the radiofrequency signal into an electrical current that

is conducted to the phrenic nerve electrodes. Electrical stimulation of the phrenic nerves leads to contraction of the diaphragm, which generates inspiration. The transmitter contains the controls for respiratory rate (frequency of electrical pulse) and tidal volume (amount of electrical voltage). DP allows full-time ventilator dependent CCHS patients to be free of PPV via tracheostomy during the day, which permits more mobility. Since the DP device is small, portable, and does not require any tubes, it allows children to participate in school and other activities without being attached to a ventilator circuit. For children who are ventilator dependent only during sleep, DP may permit tracheostomy decannulation.<sup>2</sup>

Since CCHS is a rare disease, only a few hospitals and healthcare professionals are experienced with managing CCHS patients, particularly those with diaphragm pacers. Children's Healthcare of Atlanta is the only children's hospital in the southeastern US that has a comprehensive multidisciplinary program for children with CCHS. The Diaphragm Pacing Program at Children's Healthcare of Atlanta involves collaboration of several pediatric specialists including pulmonology, general and thoracic surgery, anesthesiology, sleep medicine, cardiology, otolaryngology, critical care medicine, and genetics. This successful program has attracted CCHS patients from Georgia, surrounding southern states and as far north as Maryland. With early diagnosis and advances in home ventilation and monitoring strategies, most CCHS

# Forgetting to Breathe: CCHS

children are now surviving into adulthood, seeking higher education, and having families of their own.

For more information on the Diaphragm Pacing Program, please visit <a href="mailto:choa.org/medical-services/pulmonology/diaphragm-pacing">choa.org/medical-services/pulmonology/diaphragm-pacing</a>

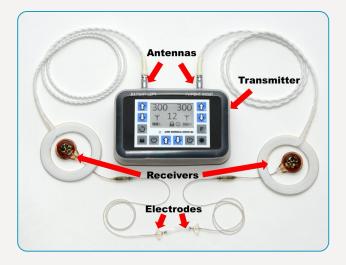


Figure 1: Diaphragm pacemaker system (Image provided by Avery biomedical devices)

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# Fall Photo Review



This year's Chapter Award honorees were recognized during the Fall Meeting in Atlanta on October 11, 2019. (I to r) Chapter Vice President Hugo Scornik, MD; Outstanding Achievement: Lynette Wilson-Phillips, MD for Kids Doc on Wheels; Young Physician Award: Katherine F. Duncan, MD & Sylvia M. Washington, MD; Denmark Lifetime Achievement: Harry L. Keyserling, MD; Chapter President Terri McFadden, MD; Georgia Dept. of Public Health Award: Jennifer Zubler, MD; Legislator of the Year: Rep. Sharon Cooper; and Chapter Executive Director: Rick Ward, CAE.



The recipient of the 2019 *Friend of Children Award* was the namesake of Beverly Knight Olson Children's Hospital Navicent Health, Beverly Knight Olson. She is joined here by Edward Clark, MD, Professor & Chair of the Department of Pediatrics at Mercer University School of Medicine.



The winners of this year's Pediatric Resident Jeopardy, (from left) Victor Ayulo, MD & Emily Nonnemacher, MD from Mercer-Macon are pictured here with Judson Miller, MD (center) and the contestants from Memorial-Savannah & Morehouse.

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# Fall Photo Review (Continued)



The Georgia Pediatric Practice Managers and Nurses Associations (P&N) met at the Cobb Energy Center in Atlanta on November 1st. The meeting brought together 127 nurses, practice managers and other office staff for a day of learning.



The P&N meeting featured a presentation on Office Emergencies by Tim Reeves (center) and Jaina Carnes, BSN (second from right). They are pictured here with Cheryl Kendall, MD (left) and her practice manager Elizabeth Taylor Pierre (right).



Chapter President Dr. Terri McFadden attended annual mtg of Ga. pediatric dentists in January and is pictured here with their leadership: Drs. Jennifer Wells, Cara DeLeon, Janice Lee & John Taylor. Wonderful colleagues in children's oral health!



During the AAP National Conference in October in New Orleans, Sally Goza, MD, FAAP of Fayetteville, was installed as President of the AAP. She is joined here by (1 to r) Rick Ward, Fozia Eskew, Terri McFadden, MD, Hugo Scornik, MD, Claire Eden, IBCLC, and Sarah Milla, MD. Congrats Sally!

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# Looking Ahead:



- May 15, 2020
   Georgia Pediatric Nurses & Practice
   Managers Association Spring Meeting,
   Macon Marriott
- April 22, 2020
  The Jim Soapes Charity Golf Classic
  Benefitting the Pediatric Foundation of Georgia
  Cherokee Run Golf Club, Conyers
- June 17-20, 2020 Pediatrics by the Sea Summer CME Meeting The Ritz Carlton, Amelia Island, FL

The Georgia Pediatrician is the newsletter of the Georgia Chapter/American Academy of Pediatrics Editor: Alice Little Caldwell, MD | Email: acaldwell@augusta.edu



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Visit the Chapter Website for details on Chapter events. www.GAaap.org
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