



# VIRGINIA • PEDIATRICS

American Academy of Pediatrics • Virginia Chapter

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## Pediatricians Focus on Gun Safety

Barbara Boardman, MD

Members of the Virginia AAP chapter mobilized in response to Virginia Governor Ralph Northam's planned special session on gun violence control which was scheduled to start Tuesday July 9. Previously, the legislative committee had polled chapter members and found that 48% of respondents listed firearm injury as a top priority.

Pediatric concerns about this are based on data that indicates that preventable childhood deaths and injury occur when firearms are not regulated. For Virginia, in 2017, according to CDC data, firearms were the number one cause of pediatric injury related deaths we lost 87 children in that year. Nationally, firearms are the third leading cause of death for children ages 1 to 17 years with 74 children injured or killed daily.<sup>(1)</sup>

Suicide deaths by firearm are an area of particular concern for those who care for impulsive adolescents. Suicide is the second leading cause of death for ages 10 to 19. Firearms are used in 40% of suicides with a 90% mortality rate. Access to firearms often decides the devastating outcome.<sup>(2,3,4)</sup>



Pictured above: Barbara Boardman, MD VA-AAP Legislative Committee member (left) with Representative Jennifer Wexton (D-VA), Christian Heyne and David Chipman at town meeting.

In preparation for the special session a letter to the editor was placed in a special section on gun violence prevention in the Richmond Times Dispatch. The letter was co-authored by Chapter president Sandy Chung, MD and Inova pediatric resident Rachel Knuth. The article advocated for 'common sense policies to help all children of the commonwealth live happy health lives,' but did not advocate for specific legislation.

Virginia chapter members were also asked to represent our concerns on various panels addressing this issue. Barbara Boardman, MD was a panelist for a town hall run by Representative Jennifer Wexton D-VA (US 10th congressional district.) She also was a panelist at a presentation for Virginia's Secretary of Health Dr. Daniel Carey in Alexandria. (photo Wexton twitter site) Lauren Morea, MD represented the AAP at a well attended panel run by Dr. Carey in Woodbridge. (photo) When interviewed about the experience of being on panels related both physicians felt there were strong concerns from the audience. Never-the-less, on the panels, the AAP panelists emphasized factual scientific data on child risk. They highlighted the role of pediatricians, who focus on asking about the practice of securing firearms, teaching about safe parental behavior and recognizing the risk of suicide.

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# President's • MESSAGE

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**Sandy L. Chung, MD, FAAP, FACHE**

President Virginia Chapter, American Academy of Pediatrics

"He's had a fever for the last two days and isn't sleeping well at night," a mom explained to me worriedly. "He has had a cold for week and I thought it was getting better. I don't know what's wrong with him!"

I look at the child sitting on her lap and the three-year old boy looks like he doesn't feel well. But he is comfortably snuggled on mom's lap in their living room in his PJ's holding his stuffed bear. He doesn't seem fazed by the fact that his doctor is talking to them from mom's phone screen.

"Can you please use the device so that I can check his ears?" I ask mom, controlling her device camera remotely and turning on the feature that works as an otoscope. I examine his ears and take video of his tympanic membranes. Then I give mom instruction on how to hold the device while I listen to his heart and lungs, measure his temperature, and examine his throat. I check his skin for rashes, and observe that he is alert and does not appear to be in distress.

Indeed, when I freeze-frame the otoscope video to get a good look at his tympanic membranes, he has a right otitis media. I let her know and then I am able to electronically prescribe the antibiotics to her pharmacy using my electronic medical record. After we disconnect our telemedicine visit, I document my note, send her written instructions, and bill for the visit all through our electronic medical record.

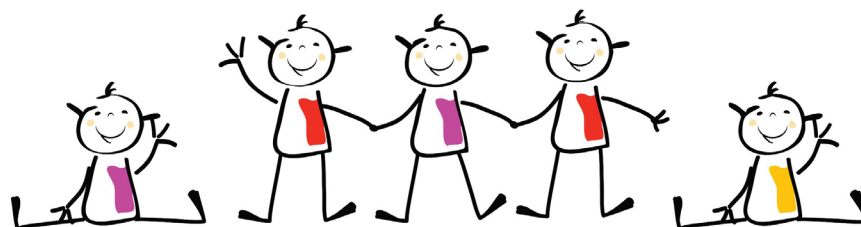
Then I click on the next patient in the virtual waiting room and continue with the next visit.

Telemedicine is here already and while it is not used as much in children as it is in adults just yet, the utilization of telemedicine in pediatrics is on the rise. Devices such as the one I am using in with my patients make the visit more palatable to pediatricians since much of a physical exam can be done remotely. While it can be a bit time-consuming to get the technology functional (as to be expected in early generation devices of any kind), the technology will only get easier to use, cheaper to access, and convenient to obtain.

The world of right-here right-now business models in practically every industry has enabled and trained patients to want super convenient everything, including health care. Controversies over the quality of care by telemedicine exist. However, even in face-to-face care, there is no question that some patients accept what some pediatricians would consider less optimal care in exchange for the highly desirable convenience factor. Often, patients do not realize that there may be a difference in the quality of care delivered to children by non-pediatricians or non-pediatric trained providers.

So, how to survive this evolving rapid, convenient care landscape? I would urge you to consider alternate means of care delivery where we come to the patient instead of the patient coming to us. We should learn from the retail-based clinic evolution that the "just say no" approach is not going to work. It is our obligation to protect the care of children and find ways to manage the way that convenient care is delivered so that children are safe. Consider telemedicine, home visits, school based care, and so on. The possibilities are endless. No matter what modality that care is delivered by, it is our responsibility to protect our patients so that they receive the highest quality, evidence-based pediatric care. Who knows? One of them may grow up and develop the next disruption in health care that will make us disease free, live forever, and make healthcare affordable for all!

Sandy Chung, VA-AAP President | [schung@fairfaxpeds.com](mailto:schung@fairfaxpeds.com)



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## PEDIATRICIANS FOCUS ON GUN SAFETY



*Pictured above: Lauren Moreau, DO, VA-AAP Gun Safety Champion and Daniel Carey, MD Virginia Secretary of Health at panel discussion in Woodbridge Virginia.*

While the Virginia chapter was well prepared for the session; the legislative response was short and disappointing. The governor had presented bills designed to limit inappropriate access and decrease lethal outcome. Some bills such as extreme risk protection orders have had national support from Republicans such as U.S. Attorney General William Barr. Never-the-less, the session closed in less than two hours deferring all consideration until after the November election.

The Virginia Chapter of the AAP is going to need your help! Please keep your eye out for Member Alerts with more information on our Advocacy Day in Richmond on January 22, 2020, where pediatricians go to Richmond and meet with legislators to advocate for pediatric issues.

1. data from Center for Disease Control WISCARS
2. Am J Pub Health 2013 jan 103.1;27-31
3. Bilsen, J Suicide and Youth Risk Factors, Front Psychiatry, 2018, 9;540
4. National Vital statistics Reports; v67#4 Jun 1, 2018; Recent increases in Injury Mortality Among Children and Adolescents aged 10-19 Years

## Congenital Muscular Torticollis

Kelli England, PhD

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Congenital muscular torticollis (CMT) is a condition in which the sternocleidomastoid (SCM) muscle is shortened on the involved side, leading to an ipsilateral tilt of the head and a contralateral rotation of the face and chin. It is a finding, not a specific diagnosis, is relatively common and is known by many names including wry neck, and loxia. It is commonly related to intrauterine positioning and often resolves with skilled physical therapy along with caregiver education on a daily home stretching program.

It is important to identify any underlying craniocervical vertebral anomalies or ocular abnormalities such as strabismus or congenital nystagmus that may be contributing to the abnormal head positioning prior to initiating therapy. Other nonmuscular causes of torticollis can include Sandifer's syndrome resulting from gastroesophageal reflux, neural axis abnormalities, and benign paroxysmal torticollis.

Previous standards of care recommended imaging of the cervical spine prior to performing active stretches to rule out concerning pathology. Treatment protocols have been updated to reflect recent recommendations based on reducing radiation in children and no longer recommend imaging unless there is concern for underlying craniocervical vertebral anomalies or lack of improvement with conservative treatment.

**KEY POINT:** Hip dysplasia is associated with CMT in up to 20% of cases

Only after failure of conservative management options should interventional options including focal botulinum toxin injections, or as a last result, surgical release, be considered. Focal botulinum toxin injections can be performed to facilitate relaxation of the spastic sternocleidomastoid muscle and allow improved stretch and range of motion in patients with persistent limitations with range of motion after a trial of more conservative treatment measures.

If left untreated, congenital muscular torticollis can lead to persistent deformational plagiocephaly, progressive facial asymmetry, and these children may be at risk for later neurodevelopmental issues. Infants with CMT who are diagnosed earlier and have earlier intervention tend to have a shorter duration of rehabilitation.



# Pediatric Horner Syndrome and Oral Considerations

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**John H. Unkel MD, DDS, MPA, FAAP**

Residency Director

**Objectives:** The reader will 1) understand the indications and contraindications for azithromycin in the acute treatment of exacerbations of problematic wheeze in preschool children, and 2) adopt recent practice guidelines advising treatment with amoxicillin/clavunate in children with protracted bacterial bronchitis syndrome.

**ACGME Competencies:** Medical Knowledge and Patient Care and Procedural Skills



Horner syndrome is a disorder involving interruption of sympathetic innervation to the orbit and is classically characterized by ipsilateral miosis, ptosis, and anhydrosis. Pseudoendophthosis, pupillary dilation lag and iris heterochromia may also be present, but depending on the specific location of insult to the sympathetic pathway, any of the typical signs may not be expressed<sup>(1)</sup>. The purpose of this report is to present a review of the recent literature regarding the clinical presentation of pediatric Horner syndrome (HS). The case of a seven-year-old male who presented to our clinic with HS of unknown etiology is also described (Figure 1). Although rare, a variety of ophthalmic manifestations of intraoral anesthesia have been recorded and we specifically discuss HS secondary to intraoral anesthesia. The symptoms of HS may not have a direct impact on oral health, but should be recognized by the pediatric dentist to make appropriate referrals based on our patients' disorders of the head and neck.

HS is a result of an interruption of sympathetic innervation along a three-neuron pathway connecting the hypothalamus to the eye, via the spinal cord and sympa-

thetic cervical trunks. The first order neurons originate in the posterolateral hypothalamus and travel through the brainstem, down along the intermediolateral column of the spinal cord to synapse at the ciliospinal center of Budge-Waller between C8 and T2. Second order neurons exit the spinal column and travel superiorly along the sympathetic chain, eventually synapsing in the superior cervical ganglion between C2 and C3. Some third order neurons travel with the external carotid artery to supply innervation to the sweat glands of one half of the face. Other third order neurons travel with the internal carotid artery, through the cavernous sinus, and enter the orbit through the superior orbital fissure with long and short ciliary fibers of ophthalmic branch of the trigeminal nerve. These third order neurons innervate the superior tarsal muscle of the upper eyelid and the pupillary dilator muscle of the iris<sup>(2)</sup>.

Since sympathetic neuronal activation and signal transduction requires norepinephrine, the main method for diagnosing HS is through manipulation of norepinephrine in the neuronal synapse. Cocaine eyedrops block the reuptake of norepinephrine from the synapse, thus blocking the expected constricting mechanism of the dilator pupillae, resulting in pupil dilation. The healthy pupil will dilate while the Horner pupil, lacking a viable sympathetic pathway, will remain constricted. Following initial verification testing with cocaine drops, hydroxyamphetamine drops can be used to help localize the lesion as either preganglionic or postganglionic, i.e. whether or not the cause of HS is located before or after the nerve synapse in the superior cervical ganglion. Jeffery et al. reported that hydroxyamphetamine is less reliable in the pediatric population for lesion localization, possibly due to the changes that occur across the synapse in a damaged immature nervous system<sup>(3)</sup>. Due to the difficulty in obtaining cocaine for testing purposes, apraclonidine eyedrops can be used to cause exaggerated dilation of a Horner pupil<sup>(4)</sup>, however, Watts et al. reported complications with using apraclonidine in the pediatric population under 6 months of age, complications which have not been reported

with the use of cocaine eyedrops<sup>(5)</sup>. Clinical presentation, the patient's history, and additional imaging (MRI, ultrasound, etc.) must be used to discriminate between insult to the first order and second order neurons as there is currently no pharmacologic test for this purpose.

Regarding specific causes of HS, the lengthy, circuitous route of sympathetic innervation to the orbit allows for a long, diverse list of etiologies. Cases of HS can be categorized as either acquired or congenital. Documented causes of acquired HS include surgical interventions, trauma, neoplasm, infection, and vascular malformation. The most common cause of congenital HS is trauma during birth, though neoplasm and vascular malformations can also serve as etiologies. Despite low comparative incidence, one of the major concerns with pediatric HS is the presence of a malignant underlying causative agent. Neuroblastoma is a sympathetic nervous system tumor that almost exclusively affects the pediatric population. It is found in 1.54 children per 100,000 per year, with a prevalence of 1 in every 7000 births. These tumors are most commonly discovered before the age of 5, many during the first month of life. It is one of the most common cancers of childhood, third to brain tumors and leukemia. The tumor can arise at any point within the sympathetic nervous system, 40% occurring in the adrenal gland, 25% in the abdomen, 15% in the thoracic cavity, and 5% in both the cervical region and pelvic sympathetic ganglia. Historically, there was a call to screen the majority of cases of pediatric HS with unknown etiology for neuroblastoma, using urinary catecholamine testing and magnetic resonance imaging of the brain, neck, and chest. More recent reports show low incidence of patients with neuroblastoma who have HS as the presenting symptom, suggesting that comprehensive testing be reserved for the most suspicious cases. Criteria for suspicious cases include absence of birth trauma, surgery, or pneumonia, the presence of ptosis, miosis, and a positive cocaine screening, as well as instability during walking and other neurologic signs<sup>(6)</sup>.

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The pediatrician should be on the aware of signs of HS especially after delivery of local anesthesia and exaggerated manipulation of the head and neck at a dental office. There have been several reported cases of the onset of HS following intraoral administration of local anesthetic. Huang *et al.* presented a case where a 32 year old woman developed concomitant HS and Harlequin syndrome following block of the inferior alveolar nerve<sup>(7)</sup>. Ostergaard *et al.* reported a case where a conscious patient developed HS presumably due to prolonged cervical extension during a dental procedure<sup>(8)</sup>. That report denies that local anesthesia delivered to the middle superior alveolar nerve could have caused the HS due to the distance of the nerve from the path of sympathetic innervation to the orbit. However, Peñarrocha-Diago *et al.* presented 3 cases where patient's developed HS-like symptoms, including enophthalmos, palpebral ptosis, and miosis following posterior superior alveolar nerve blocks and subsequent anesthesia of the ciliary ganglion<sup>(9)</sup>. They posit that HS resulted after direct diffusion of local anesthetic from the pterygomaxillary fossa through the sphenomaxillary cavity to the orbit. Despite the absence of reported cases of HS onset following administration of local anesthesia in children, one can infer a similar degree of risk given the closer proximity of anatomical structures in the pediatric head and neck as well as the increased porosity of alveolar bone in children. In the case of iatrogenic HS following dental procedures, the practitioner should be able to recognizing developing symptoms, console a potentially worried patient that the effects of the local anesthetic will likely dissipate, and recommend referral in the case that symptoms do not resolve on their own.

Salivation is tightly regulated by autonomics and one could therefor posit that patients with HS would also exhibit some change in salivation rate or content due to the disruption in sympathetic innervation. Sympathetic and parasympathetic innervation stereotypically work in antagonistic fashion throughout the body, but this over simplification is not as readily demonstrated in the control of salivation. Both branches of autonomic nerves stimulate secretion and do not work in opposition to one another. Parasympathetics are most responsible for the volume of saliva secreted and more directly control secretion of mucous than do sympathetics. Sympathetics have less affect on mobilization of fluid secreted but do not inhibit fluid secretion. Both branches stimulate contraction of myoepithelial cells. Some studies have shown that in the absence of sympathetic stimulation, protein content of saliva is reduced, though stimulation of parasympathetics has also been shown to increase protein secretion substantially. Salivary gland atrophy can occur when autonomic innervation of the gland is severed or when the reflex stimulation of the glands sparked by masticatory function is inhibited through all liquid diet restrictions. This atrophy maybe reversible if autonomic innervation is not completely severed. To date there is no data to support the idea that someone with long-term symptoms of HS experiences a quantifiable difference in salivary flow or salivary content.<sup>(10)</sup>



**Figure 1.** 7-year-old male with history of congenital Horner Syndrome; Notice discernable right-sided miosis and ptosis

A 7-year-old male was referred for evaluation of dental caries by his pediatrician (figure 1). His mother reported a history of cystic hygroma on the right neck. According to the impressions for the patient's MRI and neck ultrasounds, the neck hygroma was located between the thyroid gland and the internal carotid artery. There was no available record of the cause of the patient's HS and his mother could not recall whether or not the patient had undergone any pharmacologic testing. She also stated that she was unsure if the child had experienced any anhidrosis. Upon clinical examination, anisocoria was noted, the right pupil constricted more than the left, both in the dark and in the presence of a light source. There was also discernable right upper eyelid ptosis, at rest, with a narrower palpebral fissure on the right side. The child was referred for HS evaluation. In light of the various etiologies of HS, this patient interaction provided a helpful lesson in the importance of thorough history taking and thorough head and neck examinations.

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Judy Reinhartz PhD, Research Coordinator, St Mary's Hospital Pediatric Dentistry Residency Program, Professor Emeritus, University of Texas at El Paso

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# Chapter NEWS

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On July 24, 2019, Governor Northam, along with the First Lady, came to Hampton, Virginia to announce their Early Childhood Education Initiative. As a former pediatrician and teacher (respectively), the Northam's' commitment to children is not surprising. The Governor established the Executive Leadership Team on School Readiness since only 60% of children in our Commonwealth start Kindergarten with the required skills. Thus, the issue of disparities starts very early on in childhood. Every Virginia child, regardless of background or zip code, deserves to enter school prepared to reach his or her full potential. This initiative will hopefully have decreased the disparities in access to quality early childhood care and education which contribute to disparate outcomes for children, especially for children from economically disadvantaged backgrounds, children who are English Language Learners, and children in rural or underserved areas.

The Executive Leadership Team's core objectives include:

- 1) Ensure that all at-risk three-year-olds and four-year-olds in Virginia have access to a publicly-subsidized option; and
- 2) Ensure that all publicly-funded programs that serve children birth to five are measured as quality as part of a uniform quality rating system by 2025.

The Executive Leadership Team will be co-chaired by the Chief School Readiness Officer and the Commissioner of Virginia Department of Social Services and involve representatives from the following Departments: Education, Social Services and Planning and Budget.



*Natasha Sriraman, MD, FAAP representing the Virginia Chapter, AAP(center) discussing the Early Childhood Education Initiative with Governor Ralph Northam and First Lady Pam Northam.*



Virginia Chapter President Sandy Chung was invited to a roundtable in Norfolk on July 26, 2019, with Senator Mark Warner to discuss the protection of coverage for pre-existing conditions as well as the need to address rising pharmacy costs in our country. Several statewide health organizations including advocacy groups for patients with chronic conditions, families, and health systems were present. Dr. Chung discussed with Senator Warner the importance of protecting coverage for children with chronic conditions such as asthma, diabetes, congenital conditions, and many others. "When these children grow up to be adults, we want to make sure that they are eligible for insurance. Pediatricians need to be comfortable that treating a child for chronic conditions such as asthma does not label them such that they cannot get affordable coverage as adults in the future."

# Virginia Takes the Lead in Trauma-informed Care

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Executive Director, Eastern Shore Healthy Communities

Virginia is becoming a leader in creating trauma informed and resiliency-based communities. EVMS' well-being coalition, Eastern Shore Healthy Communities, joins 22 other organizations statewide that are offering trauma-informed training and support to multiple community sectors: health, education, social services, the courts, faith communities and others. This paper briefly describes adverse childhood experiences (ACEs), or toxic trauma, its relationship to illnesses and behavioral issues and its biological effects, what communities can do to reduce and prevent the impact of toxic trauma in children, teens and adults, and where clinicians can go to connect with Virginia trauma-informed community networks.

The scientific evidence from the ACEs study strongly associates adverse childhood experiences (ACEs), or toxic trauma, with the most prevalent diseases, behavioral disorders and social disorders.<sup>1</sup> The issues are larger and far more complex than any one sector can address alone making childhood toxic trauma and its adult outcomes a unique public health challenge. Resources are available to communities and to sectors within wishing to become more trauma informed and resiliency-based.

The Adverse Childhood Experiences (ACE) Study found a dose-response relationship between the number of categories of ACEs experienced and the number and severity of both illness risk factors and psychosocial/behavioral problems (i.e., smoking, obesity, physical inactivity, depression, suicide attempts, alcoholism, drug abuse, sexual promiscuity, and sexually transmitted diseases) and serious disease or other physical health problems (i.e. heart disease, cancer, stroke, chronic bronchitis, COPD, chronic pain, diabetes, hepatitis, and skeletal fractures) as well as health care utilization—especially rates of prescription pharmaceuticals used to treat these conditions.<sup>2,3,4,5</sup>



The ACEs study is a research initiative focusing on the role of childhood adversity, including violence and abuse and their relationship to long-term health. It describes prevalence and effects of 10 categories of ACEs including: childhood abuse (emotional, physical, or sexual), childhood neglect (emotional or physical), and household dysfunction (witnessing domestic violence, substance abuse, mental illness, incarceration, or separation and divorce) on adult well-being throughout the lifespan.

ACEs in childhood affect later mental and physical health and may be the result to changes in

the brain, endocrine and immune systems.<sup>6,7</sup> As a result of maltreatment children undergo chronic elevated levels of cortisol, a response to stress that is tolerated in the short-term, but may become detrimental to health under sustained periods of stress. Children and adults exposed to early and sustained maltreatment show elevated inflammation levels.<sup>8,9</sup> Chronically elevated inflammation levels contribute to the pathophysiology of several chronic conditions, for example, cardiovascular disease and type 2 diabetes.<sup>10,11</sup> Leptin, which acts as both a cytokine and a hormone, influences inflammation regulation and energy balance, is blunted in the face of increasing levels of physiological stimuli, inflammation, and adiposity, affecting brain development and mental health.<sup>12</sup> Also, maltreatment in childhood is linked to structural and functional brain differences.<sup>13</sup> Current research suggests that child maltreatment is associated with alterations in brain regions that may have profound negative effects on executive function, attention, memory, sequencing, planning, and visual-spatial function.<sup>14</sup>

**TO LEARN MORE ABOUT  
A TRAUMA-INFORMED COMMUNITY NETWORK NEAR YOU,**  
Voices for Virginia's Children has created a helpful site at  
<https://vakids.org/trauma-informed-va/trauma-informed-community-networks>

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ACEs are common, 41 percent of Virginia's children have experienced one ACE, and 19 percent have experienced two or more.<sup>15</sup> Resiliency is key to achieving functional status and well-being. Resilience occurs when a child or adult is immersed in positive influences, such as supportive relationships, and are protected from re-traumatization across multiple ecological systems.<sup>16,17</sup>

While logic suggests that children and adults would benefit most from mental and behavioral health therapies, it is important to note that the ubiquity of ACEs in the population may overwhelm the mental health system. Also, families often experience multiple generations of toxic trauma and for them, dysfunction is the normal state of affairs. They don't know they need help. The following is a brief outline of key environments and ways principal figures in those environments can play a supporting role in creating resilience and well-being in those impacted by toxic trauma.

- **Home.** Parents are key to helping their children develop resiliency skills within the home. When parents themselves are impacted by toxic trauma, they too, may need support from clinicians.
- **Medical settings.** Pediatricians can empower themselves with skills and resources needed to help parents understand ACEs, their causes, and ways to improve the healthy development of both themselves and their children. Gillespie and Pettersen describe a useful protocol developed at The Children's Clinic in Portland, Oregon.<sup>18</sup> Nadine Burke-Harris' Ted Talk<sup>19</sup> and her book *The Deepest Well: Healing the Long-Term Effects of Childhood Adversity* are also instructive. Other healthcare providers, including family practitioners, internal medicine specialists and obstetric providers can encourage family engagement in professional services that enhance protective factors and resources already within the child's natural environment and encourage expecting mothers and their partners to begin accessing positive support structures for themselves and their unborn child.
- **Behavioral Health settings.** Keeshin, Olafson and Cohen (2015) suggest that Trauma Focused Cognitive Behavioral Therapy demonstrates the greatest evidence base for treatment for several toxic traumatic injuries, among many other modalities described.<sup>20</sup>
- **Educational settings.** The American Psychological Association offers a Resilience Guide for Parents and Teachers<sup>21</sup> and the Centers for Disease Control and Prevention offers an online guide: *School Connectedness: Strategies for Increasing Protective Factors Among Youth* (2009)<sup>22</sup>
- **Child and Family Serving Programs.** All other sectors, like social services, court services and faith communities can join in a network with parents, medical and mental health providers, and schools to collaboratively use the best available science to support colleagues, clients and children within a network of trauma-informed and resiliency-based services that "infuse and sustain trauma awareness, knowledge, and skills into their organizational cultures, practices and policies."<sup>23</sup>

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## Newborn Screening Expands: What you need to know

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### Introduction

As of January 1, 2019, the Commonwealth of Virginia added two additional diseases to its newborn screen, Pompe disease and Mucopolysaccharidosis type I. Considered to be one of the great public health achievements in history, newborn screening began in the 1960s after Dr. Robert Guthrie developed a blood test to screen for phenylketonuria. In the nearly 60 years since that time, many more disorders have been added to the newborn screen. While the actual number of disorders varies in each state, a core minimum panel of 35 genetic, endocrine, and hematologic disorders is tested in the first two days of life. Pompe disease and MPS1 are the most recent additions to the state's screen, and the state is in the process of adding two more (spinal muscular atrophy and x-linked adrenoleukodystrophy). Disorders selected for addition to the newborn screen must have available treatments that significantly alter the morbidity and mortality of affected individuals. Here, we review both Pompe disease and MPS1, the laboratory tests used to evaluate these disorders, and the pediatrician's role in the newborn screening process for their patients.



**What is Pompe disease?**

Pompe disease (Glycogen storage disease type 2) is an autosomal recessive inborn error of metabolism caused by deficiency of the enzyme acid alpha-glucosidase (GAA). GAA resides in the lysosomes of the cell and catalyzes the breakdown of glycogen into glucose. Deficiency of this enzyme results in glycogen accumulation and tissue damage, primarily affecting skeletal and cardiac muscle. Pompe disease is typically classified into infantile-onset Pompe disease (IOPD) or late-onset Pompe disease (LOPD) depending on the age of onset, severity, and rate of progression. IOPD presents before 12 months of age (typically first few months of life) with hypotonia, generalized muscle weakness, feeding difficulties, failure to thrive, respiratory distress and hypertrophic cardiomyopathy. Without treatment, IOPD will result in death in the first year or two of life due to cardiopulmonary failure. LOPD typically presents after the 1st year of life. These individuals may have proximal muscle weakness and respiratory insufficiency without clinically significant cardiac involvement. The incidence of Pompe disease varies depending on ethnicity and geographic regions. For example, it is estimated to be 1:14,000 in African Americans but 1:100,000 in individuals of European ancestry. Enzyme replacement therapy is available for patients with Pompe disease with early treatment having significant improvements in patients with IOPD. It has also been shown that patients with LOPD have also had improvements although the timing of initiation of ERT is unclear in infants with LOPD ascertained by newborn screening.

**How is Pompe disease screened?**

In Virginia, Pompe disease is screened with a dried blood spot measuring the GAA enzyme activity level. If that level is critically low ( $< 0.625 \mu\text{mol/L/hr}$  if less than two days of age, and  $< 5.31 \text{ mol/L/hr}$  if between 2d and 1 week of age), the Newborn Screening Lab proceeds with molecular testing. Molecular testing can indicate pathogenic variants, variants of unclear clinical significance, and benign variants. A diagnosis of Pompe disease can be made with a critically low enzyme activity level and two pathogenic variants. Further evaluation is necessary if there is critically low enzyme level with non-pathogenic molecular results. One caveat is the occurrence of pseudodeficiency. Pseudodeficiency results from particular GAA gene variants that result in low GAA enzyme activity but no clinical features or diagnosis of Pompe disease. The frequency of pseudodeficiency is not fully known but it is known to be high in Asian populations. Pseudodeficiency alleles can be determined with the molecular testing performed for critical levels on newborn screening.

**What is MPS1 (Hurler syndrome)?**

Mucopolysaccharidosis type 1 is also an autosomal recessive inborn error of metabolism that results from deficiency in alpha-iduronidase (IDUA), an enzyme responsible for the breakdown of glycosaminoglycans (GAGs) in the lysosomes of the cell. Accumulation of these GAGs can result in multi-system and -organ dysfunction, including but not limited to, cardiac involvement, hearing loss, hydrocephalus, inguinal hernias, recurrent otitis media, corneal clouding, bony changes, and coarsened and dysmorphic facial features. Without treatment, individuals suffer progressive worsening of these symptoms and eventually, death. MPS1 is now divided into two subtypes, severe or attenuated MPS1, depending on the degree of residual enzyme present. Enzyme replacement therapy is available for MPS1, and individuals may also undergo hematopoietic stem cell transplant if the disease is detected early enough.

**How is MPS1 screened?**

Similar to Pompe disease, Virginia screens for MPS1 using IDUA levels on dried blood spots. If this level is critically low ( $< 4.91 \mu\text{mol/L/hr}$  if less 9 days of life), the Commonwealth reflexes to molecular testing. If the molecular testing indicates anything other than only benign variants, your local metabolic specialist may recommend further confirmatory testing. As with Pompe disease, some individuals may have low enzyme levels as a result of one or more variants of unknown significance, or may be carriers of the condition, but are not affected.

**What is the primary physician's role?**

For both disorders, the primary pediatrician will always receive notice if the screens are either abnormal or critical. Abnormal screens typically will require repeating the blood spot card. If the sample returns critical, the regional metabolic specialist is notified by the Commonwealth, and will contact the PCP to advise on the next steps.

For Pompe disease, at CHKD, we recommend that any infant with a critically low GAA enzyme level is seen by the PCP within 24 hours, and receive an echocardiogram within 5-7 days if possible. We then recommend waiting until molecular analysis is complete to determine next steps.

For MPS1, we recommend that the PCP see the child within 24 hours. As critical screens are also reflexed to molecular testing, we suggest waiting for that report before determining along with the metabolic team if further testing is necessary. The other metabolic centers throughout Virginia may have slightly different protocols or recommendations.

As with any inborn error of metabolism, it is always appropriate to call your local metabolic specialist with questions, especially if the child appears ill or you, as the PCP, have concerns. Contact information for each of the metabolic centers in the Commonwealth is listed below. Although abnormal or critical newborn screens are no doubt frightening to parents, it is important to reassure families that this is a screen, not a diagnostic tool, and that it will often detect false positives in an effort not to miss any true disease. The more educated primary providers are in regard to these diseases, the more families will feel at ease knowing the most appropriate steps are being taken to care for their child.

Parent fact sheets for both of these disorders can be found at:

[http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/Pompe\\_Education\\_Parent.pdf](http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/Pompe_Education_Parent.pdf)

[http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/MPS1\\_Education\\_Parent.pdf](http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/MPS1_Education_Parent.pdf)

Provider fact sheets can be found at:

[http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/MPS1\\_Education.pdf](http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/MPS1_Education.pdf)

[http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/Pompe\\_Education.pdf](http://www.vdh.virginia.gov/content/uploads/sites/33/2019/02/Pompe_Education.pdf)

The informational webinar available to all providers throughout the Commonwealth is available for viewing at: <https://register.gotowebinar.com/recording/3823304691012132866>

**Metabolic Centers in Virginia**

There are three metabolic centers in the Commonwealth who receive newborn screen results for each of their catchment areas. Metabolic geneticists and genetic counselors are available at each center to answer questions about the newborn screening process.

**Metabolic Diseases Program at UVA**

Division Director: William Wilson, MD  
Appointments/Referrals: 434-924-2665 or 800-251-3627 ext. 4-2665  
Fax: 434-924-1797

Clinic Location: UVA Family Medicine Primary Care Center, 4th Floor  
1215 Lee Street, Charlottesville, VA 22908  
Satellite clinics are held in Bristol, Lynchburg, and Winchester, Virginia



**Department of Human Genetics at VCU**

Division Director: Raymond Lewandowski, MD

Appointments/Referrals: 804-628-3266 or 804-628-3267

Fax: 804-828-3760 and 804-828-7094

Clinic Location: VCU Medical Center's Children's Pavilion

1001 East Marshall Street | Richmond, Virginia 23219

Satellite clinics are held in the Greater Richmond area, Fredericksburg, Petersburg, Williamsburg and South Hill

**Medical Genetics and Metabolism at Children's Hospital of The King's Daughters**

Division Director: Samantha Vergano, MD | Appointments/

Referrals: 757-668-7923 | Fax: 757-668-9724

Clinic Location: Children's Hospital of The King's Daughters, 2nd Floor | 601 Children's Lane | Norfolk, VA 23507



Children's Hospital of The King's Daughters will break ground in September on a 60-bed mental health hospital and outpatient treatment center on its main campus in Norfolk. The \$224 million project will employ 400 people and fill treatment gaps at a time when record-breaking numbers of children are in mental health crisis and searching for help. The facility is scheduled to open in 2022.

CHKD Health System's CEO and President Jim Dahling says Hampton Roads pediatricians helped the organization's board and leadership team understand the region's enormous, unmet need for pediatric mental health services. "It has truly been a groundswell from every direction," he says, "with no signs of slowing down. Pediatricians are struggling to help children with depression, anxiety, OCD, ODD and ADHD in the primary care setting. They refer more than 20 children a day to our outpatient therapy program. Another 5-10 patients arrive in our ER every day in dire need of acute inpatient, psychiatric care. We simply don't have the resources our children need." Since announcing plans for the new building, CHKD has recruited three new child psychiatrists, who are already helping to address crucial needs. "Our goal is to create a continuum of care with special emphasis on programming

that is either not available or very scarce in our region today," Dahling says. "We will offer treatment for children with eating disorders, for children between the ages of 2 and 5, for children with autism spectrum disorders and those with co-occurring medical and mental health issues. We also want to make sure that patients receive the supportive outpatient care they need to successfully reintegrate into their schools and families after an inpatient stay."

For more information, visit [CHKD.org/mentalhealth](https://CHKD.org/mentalhealth)



Barbara Kahler, MD, AAP District IV Chapter Forum Management Committee representative presented the Virginia Chapter with the 2018 Award of Chapter Excellence as well as several Special Achievement Awards for Virginia Chapter members; Samantha Ahdoot, MD, Michael Martin, MD, and Kristina Powell, MD during the AAP District IV Meeting in June. Accepting the awards were Sandy Chung, MD, VA-AAP President, Michael Martin, MD, VA-AAP Vice President, Natasha Sriraman, MD, AAP Diversity and Inclusion Champion for District IV and Jane Chappell, VA-AAP Executive Director.

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# Managing Severe Malnutrition With A Multidisciplinary Team Approach

Kyrie L. Shomaker, MD, FAAP

## The Problem

Pediatric malnutrition is defined as “an imbalance between nutrient requirement and nutrient intake, resulting in cumulative deficits of energy, protein or micronutrients that may negatively affect growth, development and other relevant outcomes.”<sup>1</sup>

Malnutrition is often considered a problem of developing countries, but is common in the US as well. Though the exact prevalence is unknown, malnutrition has been estimated to affect 14% of US adolescents, and to complicate as many as 1 in 4 pediatric hospitalizations.<sup>1</sup> Malnutrition is most commonly seen in acutely or chronically ill children, especially those with special health care needs.

Because of the significant adverse outcomes associated with malnutrition, including a 9-fold greater risk of death in severely malnourished children, exacerbation of underlying disease states, poor wound healing, and prolonged hospital stays, it is important for pediatricians to assess nutritional status, especially in high risk groups, in both the outpatient and the inpatient settings.<sup>1</sup> Screening for malnutrition includes assessment of the adequacy of protein/energy intake compared to protein/energy needs, and assessment of growth by weight, height (or length), and (where possible) mid-upper arm circumference measurement.

Malnutrition used to be defined by comparison of weight to ideal body weight, or growth crossing 2 major percentile channels over time. Now, growth charts that facilitate the comparison of the standard deviation from the mean (z score) over time are recommended. Malnutrition is identified by declining z scores or z scores greater than 1 standard deviation below the mean ( $> -1$ ). When only a single data point is available, malnutrition is identified by using the z scores for weight-for-height/length, body mass index-for-age, height/length-for-age, or mid-upper arm circumference.<sup>1</sup> When two or more data points are available, a decline in weight gain velocity (in children  $< 2$  years old), weight loss (in children 2-20 years old), or decelerating height/length z score should be used to identify malnutrition.<sup>1</sup> When is hospitalization for malnutrition appropriate?

Severe malnutrition, once identified, can be difficult to manage in the outpatient setting due to the potential for incomplete or inaccurate estimations of energy intake, the need for frequent

measurements of growth and vital signs, and often, difficulty on the part of either the patient or the family in complying with or tolerating recommended energy intake. Additionally, risks of morbidity and mortality related to the malnutrition itself, or to refeeding syndrome once treatment is initiated, escalate with the severity of the malnutrition.

Admission criteria for children with disordered eating are shown in Table 1.<sup>2</sup> Many of these markers can be extrapolated to children identified as malnourished without a formally diagnosed eating disorder, such as significant bradycardia, syncope or asymptomatic orthostatic instability, hypothermia, significant weight loss, failure to meet energy needs, and electrolyte disturbance.

Creation of the Nutritional Deficiency Protocol In 2014, after several adolescents were admitted to The Children's Hospital of The King's Daughters (CHKD) with severe malnutrition, the need for a standardized, safe approach to the treatment of malnutrition was recognized. A multidisciplinary team of pediatric hospitalists, nurses, registered dietitians, and licensed clinical social workers collaborated with local and national experts to create a protocol for the management of nutritional deficiency. Though the protocol was designed with the safety of children affected by eating disorders in mind, the intention was to create a protocol that could be used to treat any severely malnourished child, regardless of the etiology.

The Nutritional Deficiency Protocol at CHKD provides a framework for energy intake and energy expenditure that progresses based on the response of the patient to treatment. The protocol can be ordered at any point during hospitalization. The protocol triggers consultations by a registered dietitian and a member of the Behavioral Health Team, as well as specific nursing tasks such as cardiovascular monitoring, direct observation, and activity restrictions. Phase 1 of the protocol involves placement of a nasogastric tube and continuous gavage feeding for use with patients unwilling or unable to comply with recommended energy intake, whereas Phase 2 is used for patients who are able to meet their energy needs with oral intake. Though the nasogastric feeding portion of the protocol represented the most significant change from our previously established practice and was the most emotionally challenging to implement, having a collaborative and standardized approach to patients and families

in this situation has paid off. Children receive steady nutritional rehabilitation right away, and the delays in nourishment that used to occur due patient bargaining or provider inconsistency have been eliminated.

## Outcomes of Protocol

To date, 27 patients have been managed on CHKD's Nutritional Deficiency Protocol, ranging in age from 4 to 21 years, and manifesting a variety of diagnoses, including almost 50% with anorexia nervosa, as well as other eating disorders and diseases of the bowel. Patients are typically identified by their primary physician as having had weight loss, and on occasion, nearly complete food refusal or syncope.

On admission, the average BMI z score for the cohort was  $-3.3$ , with 75% of patients being formally categorized as suffering from severe malnutrition. The majority of these patients were started on Phase 1 of the protocol with continuous nasogastric feedings and frequent monitoring. Nearly half demonstrated serum phosphorus levels less than the lower limit of normal upon or shortly after admission, and were supplemented with phosphorus to prevent refeeding syndrome. Average weight gain was just over 2 pounds per week, which is within the target weight restoration goals recommended by the American Psychiatric Association to achieve improved patient outcomes.<sup>3</sup> Average length of stay for children started on the Nutritional Deficiency Protocol is almost two weeks, during which patients and families meet with dietitians and behavioral health staff on a frequent basis. Discharge disposition typically involves transfer to a regional inpatient feeding program once the medical risks have diminished, vital signs have stabilized, the child is capable of consuming the recommended meals and snacks orally, and weight gain has been established.





### How to Refer?

If you have a patient you think would benefit from CHKD's Nutritional Deficiency Protocol, call the CHKD Transfer Center (757-668-8000) to discuss your patient with the pediatric hospitalist on-call.

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**Table 1.** American Academy of Pediatrics Criteria for Inpatient Hospitalization in Eating Disorders

#### Anorexia Nervosa

Heart rate <50 beats/min daytime; <45 beats/min nighttime  
Systolic blood pressure <90 mm Hg  
Orthostatic changes in pulse (>20 beats/min) or blood pressure (>10 mm Hg)  
Arrhythmia  
Temperature <96°F  
<75% ideal body weight or ongoing weight loss despite intensive management  
Body fat <10%  
Refusal to eat  
Failure to respond to outpatient treatment

#### Bulimia Nervosa

Syncope  
Serum potassium <3.2 mmol/L  
Serum chloride <88 mmol/L  
Esophageal tears  
Cardiac arrhythmias including prolonged QTc  
Hypothermia  
Suicide risk  
Intractable vomiting  
Hematemesis  
Failure to respond to outpatient treatment

From Campbell K and Peebles R. Eating disorders in children and adolescents: State of the art review. *Pediatrics* 2014;134:582-592.

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