

VIRGINIA • PEDIATRICS American Academy of Pediatrics • Virginia Chapter

Summer | 2014

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American Academy of Pediatrics DEDICATED TO THE HEALTH OF ALL CHILDREN

Virginia Chapter About Us

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Legislative Conference in Washington D.C.

KRIS POWELL, MD, FAAP | VA-AAP DELEGATE EASTERN VIRGINIA

The legislative conference is a terrific learning experience for all pediatricians – including medical students, general practitioners, subspecialists and retirees. It is an opportunity to gather together with a group of diverse people who share a similar goal: to support pediatric priorities in our country.

This year's conference was held in Washington DC. Attendees had three full days of education with plenty of hands on experience. Discussions included how advocacy is important, how it works and what we each can do to continue this work through the rest of our careers. Although we learned about advocacy in general, there was a focus on childhood nutrition as it is an active legislative priority. Specifically, we discussed the importance of upholding the Healthy. Hunger-Free Kids Act, a bipartisan law passed by Congress in 2010. which requires the US Department of Agriculture to improve nutrition standards for the National School Lunch Program. The law requires school meals and snacks to include more whole grains, fresh fruits and vegetables while limiting added sodium and unhealthy fats. The law has been rolled out in phases and this year will be the final year for full implementation. Although the cost of school lunches have gone up, the schools that participate get reimbursed for each meal. Also, the long term savings of healthy school lunches are unmeasurable. Today, 90% of schools are meeting these requirements.

On June 11, 2014, the FY 15 House Agriculture Appropriations Bill was introduced that would require the USDA to provide waivers to schools which would allow those struggling to meet the requirements, to opt out. Specifically, if a district shows a net loss in its food service over a six month period, they can opt out. The language for "loss" is vague. The AAP is opposing the efforts to weaken the standards, arguing that it would be best to find ways to help the 10% of schools that have been unable to provide the standards and to continue to support the 90% of schools that have. Representative Sam Farr (D-California) has offered an amendment to strike the waiver provision. The outcome of this proposal is yet to be determined. We also discussed the different proposal to include white potatoes into the WIC program which the AAP obviously opposes.

Overall, the conference was a memorable experience. Health policy and advocacy are rewarding areas of pediatrics. Although we, as individuals, may feel uninfluential, advocacy groups and conferences like these, help participants realize that our voices really do matter.







President's • MESSAGE

Barbara M. Kahler, M.D.

President Virginia Chapter | American Academy of Pediatrics

It's that time again when we change over leadership. We wish to thank Biff Rees for his Leadership through interesting times. We wish him well as he continues to work with Chamber of Commerce and early childhood initiatives. We hope he continues his valuable presence on the Board.

We welcome Sam Bartle, MD, FAAP as Vice President and Sandy Chung, MD, FAAP as Secretary -Treasurer.

Now for what's new:

- 1) MOC Part B will now be part of your membership through EQUIPP
- 2) Membership Committee is looking at different ways to restructure dues
 - Buffet of benefits to choose from with dues based on your choices ie; EQUIPP, Red Book, Prep, etc.
 - Dues to be paid yearly with possible option to pay monthly
- 3) Loan consolidation help for those coming out of training
- 4) Committee on Infectious Disease- Controversial Synagis statement is due out soon.
- 5) AAP is looking for innovative ways to enhance HPV vaccine rates

These are interesting times for Pediatrics in Virginia and Nationally. We must work collaboratively to preserve our place at the table. So look for more from National and the Virginia Chapter on how we are working with others to promote the well-being of children, especially those under 3. Also how to promote resilience in our children exposed to toxic stress and poverty.

I am looking forward to working with you all during the next two years.

Sincerely,

Larbara Lat



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CHKD Center for Hemangiomas and Vascular Birthmarks

Established in 2000, the CHKD Center for Hemangiomas and Vascular Birthmarks has kept pace with the evolving management of these lesions. With the emergence of propranolol as the preferred intervention for infantile hemangiomas of functional and cosmetic significance, our doctors have participated in the development of a consensus statement guiding the initiation of this medication. We are also co-authoring the American Academy of Pediatrics Clinical Report on Infantile Hemangiomas, and we have established a clinical grading and staging system for these lesions. We have also published the first investigation of oral cavity changes in patients with port wine stains.

Staffed by CHKD Drs. Judith Williams from Pediatric Dermatology, David Darrow from Pediatric Otolaryngology, and George Hoerr from Pediatric Plastic Surgery, our vascular anomalies team treats the gamut of vascular lesions in children, including hemangiomas, port wine stains, and venous and lymphatic malformations. Additional consultants in orthopedic surgery, general surgery, head and neck surgery, and interventional radiology also participate as needed. Our physicians collaborate to design a treatment plan customized for each patient that will often involve the participation of several members of the physician team.

Due to the rapid growth and acute symptoms associated with these lesions, the team will see children with these lesions on short notice. The clinic meets twice a month at CHKD in Norfolk and every other month at the CHKD office in Newport News. Appointments can be made through Jennifer Blackburn, LPN, Coordinator for the Center for Hemangiomas and Vascular Birthmarks, at 757-668-7793.

Additional information may be found at http://www.chkd.org/Services/Vascular/.



Dates to Remember

The Donald W. Lewis Pediatric Update 2014 September 19th – 21st, 2014 Williamsburg Lodge Williamsburg, Virginia

For details contact Rosalind Jenkins 757/668-8942 or Register online: www.chkd.org/DonaldLewisPediatricUpdate

Mohsen Ziai Pediatric Conference November 7th & 8th, 2014

The Ritz Carlton Tyson's Corner, Virginia

For more information, please visit www.inova.org/pedscme or contact Marchelle D. Albertson at marchelle.albertson@inova.org

Pediatric General Assembly Day Thursday, January 29th, 2015 7:30 AM – 2:00 PM

The home base for the Pediatric General Assembly Day Hilton Garden Inn Located at 501 E. Broad Street In Richmond. This venue is a flat, 3.5 block walk from the General Assembly Building. Shuttle service will also be available. The Hilton Garden Inn offers valet parking and is convenient to several public parking lots.

For more information go to www.virginiapediatrics.org after January 1, 2015

www.virginiapediatrics.org

CHILDREN'S HOSPITAL OF THE KING'S DAUGHTERS AND THE AMERICAN ACADEMY OF PEDIATRICS, VIRGINIA CHAPTER

Present

VIRGINIA PEDIATRICS NEWSLETTER American Academy of Pediatrics – Virginia Chapter

Continuing Medical Education

This activity has been planned and implemented in accordance with the Essential Areas and policies of Medical Society of Virginia through the joint sponsorship of Children's Hospital of The King's Daughters and the American Academy of Pediatrics – Virginia Chapter.

Children's Hospital of The King's Daughters designates this enduring material for a maximum of 3.0 AMA PRA Category 1 Credit(s) ™. Physicians should only claim credit commensurate with the extent of their participation in the activity.

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How to Obtain Credit:

Review the articles on pages 5 -20. Complete the attached VA-AAP Newsletter Registration and Evaluation Form and return to the Children's Hospital of The King's Daughters, CME Office, 601 Children's Lane, Norfolk, VA 23507, or 757-668-7122. You may also visit https://www.surveymonkey.com/s/VAAAPSummer2014 and complete online. Please allow 8 weeks to receive your certificate.

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Drowning: Focus on Prevention

Rianna Evans MD Pediatric Hospitalist, CHKD Assistant Professor of Pediatrics, EVMS

Virginia offers a variety of outdoor summertime activities for children including recreational swimming in salt-water, freshwater and community swimming pools. With these activities comes an increased risk of accidental injury and drowning during the summer months.¹ Pediatricians should be knowledgeable on the medical care for patients with drowning injuries and be actively involved in the education of caregivers and the community on the prevention of drowning events.

Drowning is a leading cause of injury and the second leading cause of death in children. Approximately 70% of all drowning related visits to the Emergency Department in the United States from 2005-2009 occurred in children less than 14 years of age. While there is an increased risk in summer months related to recreational water activities, there is an under-recognized risk for drowning in shallow water such as bathtubs, which is the primary location for drowning in infants < 12 months of age. Children in low-income and urban households are at an increased risk of drowning with death rates highest in those less than 4 years of age, blacks, adolescents and males.¹

The terminology used to describe drowning events has changed in recent years to standardize populations and treatment options. The World Health Organization and World Congress on Drowning have defined drowning as "the process of experiencing respiratory impairment from submersion/immersion in liquid". As the physiologic effects from submersion in liquid may occur up to 24 hours after the submersion, the terms near-drowning, wet-drowning, dry-drowning and secondary drowning should no longer be used. Events are classified as either fatal or non-fatal, with non-fatal events further classified into those with morbidity and those without morbidity. Distinctions between the type of liquid in which the person is submersed (i.e. fresh-water versus saltwater) are also no longer made, as research has shown that there are no true physiologic differences based on the submersion liquid in patients with non-fatal drowning.²

Despite the change in terminology in the medical community, there has been increasing media presentation on the topic of "secondary drowning", prompting concerned parents to seek advice from pediatricians. This term refers to the pulmonary edema that occurs several hours after a seemingly minor drowning event. Parents can be reassured that this is a rare event and that if a child develops respiratory distress, severe cough or altered mental status within 24 hours after such a concerning event in the water, medical care should be sought.

Morbidity related to non-fatal drowning can involve respiratory compromise, neurologic sequelae, hypothermia and injuries related to objects or chemicals in the body of liquid. If present, respiratory complications are largely manifested as acute respiratory distress syndrome (ARDS) resulting from pneumonitis and wash-out of surfactant from fluid aspiration, causing alveolar collapse. The resultant hypoxemia from fluid aspiration causes end-organ ischemia including neuronal damage and cerebral edema, renal compromise from acute tubular necrosis and cardiac dysrhythmias. The key treatment focus in patients with non-fatal drowning is therefore focused on ventilation to decrease end-organ damage from ischemia.³ The aggressive management of cerebral edema and seizures can also improve neurologic outcomes; the role of therapeutic hypothermia is evolving and remains controversial, with research ongoing and use in children after cardiopulmonary arrest made on a case-by-case basis.4,5 Factors related to worse neurologic outcomes include prolonged submersion time, prolonged resuscitation time and a delay to

Objective: Participant will be able to review the epidemiology, presentation and medical care for pediatric drowning victims and to discuss prevention efforts for pediatricians. **ACGME Competencies:** Patient Care, Medical Knowledge

initiation of bystander CPR. 6,7

Prevention of drowning events involves layers of protection and education in the community. The primary preventative measure for children is adult supervision. The coined term 'touch supervision' is when an adult is within arms reach of the child and fully attentive to the child's needs, without engaging in distracting activities.² This close supervision applies to bath time as well as recreational water activities. Though touch supervision at all times is ideal, there are expected lapses in many circumstances and other measures can help promote water safety. The use of swimming lessons and water-survival skills to help prevent drowning events in young children is controversial. The American Academy of Pediatrics has classically discouraged swimming lessons in children less than 4 years of age because young children are unlikely to be developmentally ready until this age and there was a concern that swim lessons may create a false sense of safety in a high-risk population; however recent studies have suggested that swimming lessons do not increase drowning events and may decrease fatalities in some young children.⁸ With this evidence, the age to initiate swimming lessons is left to parents as an individualized decision, with the knowledge that no degree of teaching will fully prevent a drowning event.²

Safety equipment for recreational swimming areas may also decrease the risk of drowning events. All persons with inadequate swimming skills should wear a US Coast Guard approved Personal Flotation Device (PFD, lifejacket) and the use of other air-filled swimming aids should be discouraged. Particularly for young children, the PFD should be kept on when in or around recreational bodies of water as most drowning events in children occur when children are unsupervised (such as dur-

... (cont.) Drowning: Focus on Prevention

ing a family pool party). For all swimming pools (including above ground or inflatable pools) a 4-sided fence with a self-latching, self-closing gate should be used to prevent access to the pool by unsupervised children. Pool alarms and rigid pool covers may be used to supplement a 4-sided fence, but have not been proven to decrease the risk of drowning when used alone. A particular risk to young children is entrapment in vacuum drain devices and drain covers in swimming pools, therefore residential pools owners should be instructed to install safety vacuum-release systems and special drain covers. As early retrieval from submersion and initiation of CPR are crucial to improving neurologic outcomes, caregivers and pool owners should be trained in CPR and the proper use of life saving equipment.²

Pediatricians are encouraged to provide general anticipatory guidance on water safety to all families and to identify those with residential swimming pools or residence on a body of water to provide more targeted advice. In addition, we may improve safety for water activities through participation in legislative efforts to mandate safety equipment in residential and natural swimming areas and ensure proper emergency medical services are in place for children in our regions.²

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From Hungry To Healthy

The Importance of Increasing School Breakfast Participation



School breakfast plays a critical role in helping school children reach their full academic potential. This may be especially true for the 1 in 5^t who live in a household faced with food insecurity.

To help our nation's children who need to move from hungry to healthy², we are committed to increasing student participation in School Breakfast Programs.

Collectively we will work together to:

- Increase awareness of the critical impact School Breakfast Programs have on learning, nutrition security, diet quality and student health.
- Provide resources to empower schools to champion school breakfast.
- Inspire families and communities to embrace school breakfast.
- Empower children to take action to help increase access to breakfast in their schools.



(Insecutly in Novehold) WPB Childrer: Prevalence, Severity, and Household Characteristics, 2003 41 by Alista Calenzan-Jenner, William Hickall, and Hash Nand, Gonnenic Information Builetin Ho. (18) 411 (19) pp. May 2013 Topocolonately 1% of households experience very low food security where children are hunger, daip a meat, or don't eat for a whale day because of economic challenges at some time during the year.

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Food Allergy Diagnosis: Pearls and Pitfalls

Heather B. Minto, MD Kelly M. Maples, MD Children's Hospital of The King's Daughters, Department of Allergy and Immunology

Approximately 6 million or 8% of children in the United States have food allergies. Younger children are impacted most. As food allergies become more common, so does the misdiagnosis of food allergy.

Immunocap or specific IgE testing to foods is easy to order and does not require a visit to an allergist, however this test used alone can lead to many false positives, resulting in unnecessary dietary limitations and anxiety for families. she followed his doctor's advice until her son was evaluated by an allergist.

Oral food challenges performed in a physician's office prepared to treat anaphylaxis are the gold standard for food allergy diagnosis. Both allergy skin prick testing and immunocap testing, while helpful tools, have high false positive rates and can lead not only to unnecessary dietary elimination but also may lead to loss of oral tolerance to the food in question when used inappropriately.



An illustrative case concerns a 7-year-old boy who had an allergic reaction after eating peanut butter crackers. Within minutes, he developed wheezing, urticaria, and angioedema which was treated in the emergency department. When he followed up with his primary care physician, immunocap testing was ordered to a panel of multiple foods, including foods he was tolerating. He had specific IgE detected to peanut (27 kU Ab/L), but also to wheat (0.72) and milk (0.94), which he ate daily without reaction. His primary care physician told the family to remove peanut, wheat, and milk from his diet. His mother was confused since he ate wheat and milk without any problems, but

Children with atopic dermatitis often have a very elevated total IgE, which can lead to false positive rates above those of children without atopic dermatitis. The above patient reintroduced milk and wheat into his diet without complication.

Fleischer et al performed oral food challenges in children who

were avoiding foods due to positive skin prick test, food-specific IgE, or history of a reaction. Patients with a history of a life threatening reaction, a convincing history of an acute reaction in the preceding 6 to 12 months, an immunocap IgE level greater than the 95% predictive index or a large associated skin prick test did not undergo OFC. Three hundred and sixty-four oral food challenges (OFC) were performed to foods previously avoided. 89% (325) were negative. On subgroup analysis, 111 OFCs were performed in 44 children who were avoiding foods solely due to either positive food-specific IgE or skin prick test without history of reaction. Over 80% of these OFCs

Objective: Participant will be able to recognize proper food allergy diagnostic techniques. Recognize the risk for over diagnosis of food allergy. To explain the role of component testing in food allergy diagnosis **ACGME Competencies:** Patient Care; Medical Knowledge

were negative. Many children in the study were on unnecessary avoidance diets and were avoiding foods that they had never been exposed to or had previously tolerated as the result of improperly used foodspecific IgE testing. Similar results were seen in OFC's done for previous nonanaphylactic reaction, where 70% of the OFCs were negative. Overall, the majority of the OFCs in this study were negative, (89%).

If food allergy is suspected, checking an immunocap to the food in question is a reasonable approach but avoid checking immunocaps to an indiscriminate panel of foods or to any foods a child is regularly tolerating. Be sure the patient is also instructed to maintain two epinephrine autoinjectors at all times and how to use them. They must also be provided with a life-threatening allergy management plan with emphasis placed on the importance of early epinephrine administration during systemic food allergic reactions. Failure to administer epinephrine rapidly (within minutes) is a risk factor for food allergy fatality.

Many food allergies are outgrown and an allergist can follow skin testing and immnocap results over time and when safe, perform supervised oral food challenges. When interpreting immunocap results, it is important to note that the class system of reporting immunocaps is helpful in environmental allergy only and has no role in the diagnosis of food allergy. For each food allergen, a different specific IgE threshold is used to determine if a patient is eligible for an oral food challenge. Any provider performing food-specific IgE testing should be familiar with the IgE threshold for each food in question. IgG testing to foods is not diagnostic of any condition and should not be ordered. Normal individuals produce IgG to foods to which they have been exposed. Intradermal skin testing should also never be done to foods because it can cause anaphylaxis in allergic individuals and has a very high false

... (cont.) Food Allergies Diagnosis: Pearls and Pithalls

positive rate.

Newer component testing for IgE to specific portions of food proteins is now available. IgE levels to ovomucoid can be used to see which patients are eligible for a baked egg challenge. Many patients with egg allergy can safely eat baked eggs. This does not mean that they are any less allergic to nonbaked eggs, but allows a less restricted diet. Recent studies show that regular consumption of baked egg may help children outgrown egg allergy faster. Specialized peanut component testing is also available. Some patients are sensitive to a portion of the peanut protein similar to pollens (prophyllins). These children will likely have mild reactions to accidental peanut exposure. Others are allergic to portions of the peanut protein unique in nature to peanut and are at high risk for anaphylaxis with accidental exposure, regardless of the severity of previous reactions or size of skin prick wheal.

After taking a detailed dietary history, targeted food-specific IgE testing and skin prick tests are helpful in identifying those individuals with likely food allergies. However, in patients who have not experienced anaphylaxis to a particular food, sole reliance on food-specific IgE testing is not adequate to diagnose food allergy. Oral food challenges remain the most accurate way to diagnose food allergy in those patients without history of anaphylaxis.

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5. Bock SA, MunozFurlong A, Sampson HA. Fatalities due to anaphylactic reactions to foods. J Allergy Clin Immunol. 2001; 107(1): 1913. Is My Child's Brain Going to be Hurt by Anesthesia? Anesthesia and Neurotoxicity in the Pediatric Patient; Overview and Current State

Objective: Participants will be able to describe the concerns surrounding this evolving issue of anesthesia. Discuss the current evidence from animal and human studies regarding this topic. Recognize this issue's potential impact on the practice of health care professionals involved in the provision of pediatric anesthesia or sedation and those whose patients need procedures requiring anesthesia or sedation. Explain the current state of knowledge of anesthesia neurotoxicity on the immature human brain to our patients' inquiring caregivers as stated by published professional consensus statements.

ACGME Competencies: Patient Care, Medical Knowledge, Practice-based Learning and Improvement, Interpersonal Communication Skills, Professionalism, Systems-based Practice Knowledge

Introduction

In recent years there has been growing evidence from animal studies that commonly used anesthetics cause apoptosis in the immature animal brain and are associated with long term learning disabilities in the affected animals. In addition, some human epidemiological studies have also questioned the safety of anesthetics in the developing human brain and found associations, not causality, of learning and neurobehavioral deficits in children after exposure to anesthesia and surgery at an early age.

With over 4 million anesthetics provided to children yearly in the US alone with many surgeries being critical to the child's wellbeing, this issue has led to great concern and much focused research efforts on the part of the international pediatric anesthesiology community. New information resulting from sizable ongoing research makes this a rapidly changing field. Since many healthcare professionals administer pediatric sedation and anesthesia in varied settings outside the operating room such as emergency departments, radiology suites, dental offices, neonatal and pediatric intensive care units (NICU and PICU); this issue can have major impact on practice decisions by anesthesia and sedation providers and by specialists whose patients require sedation or anesthesia for diagnostic or therapeutic procedures.

Historically, anesthesia and analgesia for neonates and small infants were thought to be unsafe due to detrimental drug effects on immature organs and lack of age appropriate equipment, or unnecessary with the

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Destiny Chau MD Associate Professor Miriam Schoepf MD Assistant Professor Children's Hospital of The King's Daughters Division of Pediatric Anesthesiology



argument that certain pain pathways were not yet developed in these young patients. Subsequent research revealed that neonatal rat pups have significant stress responses to repetitive painful stimulation with associated adverse behavioral outcomes including neuronal cell death. Therefore, untreated pain leads to neurotoxicity on the developing nervous system while adequate analgesia confers neuroprotection. Withholding anesthesia and analgesia from our youngest patients during noxious stimulation is unacceptable as the failure to blunt stress responses and treat pain is clearly associated with harm.^{1,2,3,4}

As reports of anesthetic neurotoxicity have reached the lay press and media, a growing number of well-informed parents are anxiously asking about this topic. What do we say? First, let's review the evidence.

Current State of Knowledge - Animal Studies

A leading researcher at the University of Virginia, Dr. Jevtovic-Todorovic, published a landmark paper in 2003 describing widespread apoptosis in the developing rodent brain following exposure to routine anesthetics with subsequent maladaptive effects.⁵ Since then, over 300 studies and abstracts on different animal species ranging from mice, rats, chicks, guinea pigs, swine and sheep to non-human primates have replicated these findings. Intrauterine anesthetic exposure has also been linked to neuronal apoptosis in rhesus monkeys^{6,7}, while pregnant rat exposure has been shown to lead to persistent neurobehavioral deficits in their offspring.⁸ Physiologic apoptosis, or

continued from page 8...

naturally occurring programmed cell death, in the immature brain is a normal process necessary for healthy nervous system development. This contrasts to the accelerated widespread apoptosis and neuronal degeneration observed in these animal studies which coupled with long-term neurocognitive abnormalities is evidence of pathologic processes. All implicated anesthetics fall into the NMDA antagonist and/or GABA agonist families which include sevoflurane, desflurane, isoflurane, enflurane, halothane, nitrous oxide, xenon, ketamine, fentanyl, morphine, propofol, chloral hydrate, pentobarbital, diazepam and midazolam. The list covers most agents commonly used to provide sedation and anesthesia not only in the operating room but also in the NICU, PICU and other sites. Combining anesthetic agents of differing mechanisms may result in greater neurotoxicity than each when given alone. Such induced neurotoxic effects on the developing animal brain seem to depend on both anesthetic dose and length of exposure above thresholds that are currently undefined. Existing adult research reporting neuroprotective effects of anesthetics adds to the confusion in this topic.9 Recent data suggest that exposure during vulnerable periods of maximal synaptogenesis is critical for neurotoxicity while exposure past this window may confer neuroprotection.¹⁰ This vulnerability is associated with cell type and age.^{11,12} Histological changes are not necessarily followed by clinical changes 13 with the reverse apparently also being true. Studies that have also included noxious stimulation to simulate surgical stress and pain have shown conflicting results.^{14,15,16} Simplified theories for mechanisms of anesthetic neurotoxicity include: 1) Suppression of neurotrophic signaling pathways, 2) suppression of synaptic activity, 3) suppression of prosurvival extracellular signaling pathways, 4) anesthetic associated pro-inflammatory effects, 5) induction of seizure activity leading to neurotoxicity and 6) mitochondria effects.17,18,19,20,21

Research on mitigating strategies have shown the following agents as promising : the $\alpha 2$ adrenergic agonists dexmedetomidine and clonidine, ketorolac, hypothermia, lithium, melatonin, B-estradiol, L-carnitine, xestispongin-C, bumetamide and erythropoietin.^{22,23,24,25} Early studies showed xenon to be neuroprotective but new studies indicate neurotoxicity at adequate clinical doses.²⁶ Also, environmental enrichment after anesthetic exposure seems to improve neurobehavioral and neurocognitive function.¹⁴

Current State of Knowledge -Human Studies

Human data on anesthetic neurotoxicity is far less conclusive than animal data. Currently available human studies are scarce (about a dozen) and are based on retrospective or observational studies which do not address causality. Some report positive associations of adverse neurological effects with surgery and anesthesia exposure while others did not find such effects. The associations found are usually weak. Examples of the studies include: 1) Patients with congenital heart disease after heart surgery and cardiopulmonary bypass; the findings report abnormal motor and cognitive development through adolescence. Limitations include the difficulty separating the effects of repeated anesthetic exposures to the effects of surgical interventions, cardiopulmonary bypass, hypothermia, preexistence of brain abnormalities and other confounding variables.^{27,28} On the other hand, Guerra and colleagues²⁹ did not find such association in a similar study. 2) An Australian cohort of 2287 controls and 321 patients with one exposure to surgery and anesthesia found a risk ratio of 1.7-2.1 for receptive and expressive language deficits for exposure at age 3 years or younger. No deficits were found in vocabulary, behavior and motor functions.³⁰ 3) In a Danish birth cohort of 14575 controls and 2689 patients undergoing inguinal hernia repair. No test score difference was found on an academic achievement examination. A bias is that the exposed group was less likely to sit for the exam.³¹ 4) Bartels et al³² studied 1142 monozygotic concordant and discordant twin pairs where 1 sibling had an anesthetic exposure prior to 3 years old. Educational achievement tests indicate the males scored significantly lower than the population norm while the females were non-significantly lower; however, no differences were seen for discordant twins. 5) A birth cohort of 4764 controls and 593 patients exposed to anesthesia and surgery before 4 years old found by standardized aptitude school district testing that a single exposure was

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not significant for development of learning disabilities but cumulative exposures and increased anesthesia duration increased risks. Limitations include the 1976 -1982 study period when obsolete anesthetic agents such as halothane were common and pulse oximetry was not yet available for perioperative monitoring.³³ 6) A sibling cohort from 1999-2005 consisting of 10146 controls and 304 exposed to anesthesia and surgery prior to 3 years old. Results found that the exposed group had 60% greater risk of developmental and behavioral disorders; however, tighter analysis did not find this effect.³⁴

The variation in these results can be largely accounted for by the limitations and presence of confounding variables inherent to the retrospective and observational nature of these studies. It is also necessary to emphasize that the differing outcome measures and end points used by the studies contribute to the variability of the findings.^{35,36}

How Are Researchers Currently Addressing This Issue?

Not enough human data is currently available and extrapolation of animal data to the human species is plagued with pitfalls such as issues of equivalence of anesthetic dosing, length of exposure and developmental and neurophysiologic timing differences across species. Therefore, several major initiatives are currently underway to move human research forward:

SmartTots (Strategies for Mitigating Anesthesia-Related neuroToxicity in Tots) is a research initiative of the International Anesthesia Research Society (IARS) which has partnered with the US Food and Drug Administration (FDA) in a multi-year collaborative effort to fund research seeking answers to the following key questions:

1. What is the spectrum of general anesthetic agents, sedatives, surgical procedures, and/or opiates that cause developmental neurotoxicity? What are the doses, durations, and frequencies of exposure? What are the most vulnerable periods of development?

2. Are there short- and long-term neurocognitive, emotional, behavioral, and/or social outcomes resulting from exposure to anesthetic agents?

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3. What approaches can be taken to prevent or mitigate developmental anesthetic neurotoxicity?

The large, multi-center PANDA (Pediatric Anesthesia & Neurodevelopment Assessment) project studies children exposed to anesthesia for inguinal hernia surgery before age three and compares them with non-exposed children while following their neurodevelopment and cognitive functions between ages 8-15 years old.

The MASK study (Mayo Safety in Kids) is a population-based study in collaboration with the National Center for Toxicological Research of long-term cognitive development in a large cohort of children in Rochester, MN. It compares children's performance without anesthetic exposure to those with one or more exposures prior to age three.

The GAS (General Anesthesia Spinal Anesthesia Study) study is an international, multi-site, randomized controlled trial examining the long-term effects of spinal versus general anesthesia used in newborns undergoing hernia surgery with regard to neurodevelopmental outcomes at ages 2 and 5 and the incidence of apnea in the postoperative period after regional versus general anesthesia.

So What Do We, Pediatric Practitioners, Do In The Meantime?

Based on the available data a change in current practice for administration of sedatives and anesthetics is not warranted. Delaying necessary procedure or surgery appears more risky.

How Do We Answer Parents' Questions Regarding Anesthetic Safety And Neurotoxicity?

The following widely accepted statements can be used as guidelines when speaking with parents:

SmartTots' consensus statement (www. smarttots.org/resources/consensus.html) released on December 2012 regarding the use of anesthetics and sedatives in children states that "additional research is urgently needed...it would be unethical to withhold sedation and anesthesia when necessary... Healthcare providers should discuss with parents and other caretakers the risks and benefits of procedures requiring anesthetics or sedatives, as well as the known health risks of not treating certain conditions... Physicians should remain informed on developments in this area, but recognize that current anesthetics and sedatives are necessary for infants and children who require surgery or other painful and stressful procedures".

This statement has been endorsed by the FDA, IARS, American Academy of Pediatrics, Society of Pediatric Anesthesia, Society for Neuroscience in Anesthesiology and Critical Care, American Society of Anesthesiologists and European Society of Anesthesiologists.

The FDA released in 2013 the latest consumer update on anesthesia safety on the young brain (http://www.fda.gov/forconsumers/consumerupdates/ucm364078. html) and states that "caretakers must talk to their pediatrician or other health care professionals about the risks and benefits of procedures requiring anesthetics and weigh them against the known risks of not treating certain conditions."

When responding to caregivers asking about anesthetic neurotoxicity, we need to put the risks and benefits in the context of what we currently know and do what we do every day for all our patients -- namely what we perceive is best for the child. Once the intervention or surgical procedure is deemed essential for the child's wellbeing and delaying the procedure would create more risk, we need to continue to provide adequate anesthesia and analgesia to minimize the wellknown risks induced by surgical procedures and associated pain on our young patients.

In summary, although the current human data suggest an association between anesthetic and sedative agents and neurodevelopmental deficits, it does not indicate causality. On the same token, there is also insufficient data to dismiss these concerns. The current evidence is insufficient to warrant a change in anesthesia and sedation practice. More risk and harm would be incurred by attempting to minimize risks that are currently undefined. With that in mind, we challenge you to continue to stay tuned regarding this important evolving topic.

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Complement Inhibitors and Their Potential to Moderate Ischemia-Reperfusion

Injury in Patients With Sickle Cell Disease

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Objective: Participants will be able to review the physiology of Galactosemia, as well as the clinical presentation of critical newborns with the disorder, and recall the necessary steps to follow after receiving a critical or abnormal newborn screen for galactosemia. **ACGME Competencies:** Patient Care, Medical Knowledge.



September is National Sickle Cell Awareness month in the U.S. In Virginia, newborn screening identifies about 75 newborns with sickle

cell disease (SCD) every year. About 4,500 Virginians are living with SCD. Increasing grant support and work in overcoming barriers to health care among patients with SCD is occurring in collaboration with Virginia Commonwealth University (VCU), Eastern Virginia Medical School (EVMS). Children's Hospital of The King's Daughters (CHKD) and the Virginia Department of Health. Based in Norfolk, EVMS serves as one of the two largest sickle cell programs in the state with more than 500 children with SCD cared for by CHKD. In recent years, new evidence suggests that inflammation plays a major role in mediating host tissue damage for patients with SCD.

A critical component of the host defense, the complement system is a potent inflammatory cascade in humans. It is mediated through 3 pathways: the classical, alternative and mannose binding lectin pathways. Discovered first, the classical pathway of the complement system is mediated by the C1 molecule. It is comprised of C1q, a pattern recognition molecule which detects surface bound antibodies and is associated with 2 serine proteases, C1r and C1s.^[1] The immunoglobulin molecules: pentameric IgM and clustered monomeric IgG are recognized by C1 which in turn cause activation of C4 and C2 to form the classical pathway C3 convertase (C4b2a), another protease complex. C3 and terminal cascade activation lead to the generation of inflammatory anaphylatoxins (C3a and C5a) and the lytic membrane attack complex (MAC). C1g also plays additional roles in clearing cellular debris, immune complexes and apoptotic cells [2, 3] through recognition of PAMPs (pathogenassociated molecular patterns) and DAMPs (damage associated molecular patterns). The complement system comes equipped with various regulatory proteins to limit unwarranted host tissue damage. However, deficiency of these regulatory proteins or aberrant activation of the system causes significant host tissue damage and has been implicated in the pathogenesis of many inflammatory diseases, including systemic lupus erythematosus (SLE), acute intravascular hemolytic transfusion reaction (AIHTR). hemolytic disease of the newborn, serum sickness and ischemia reperfusion injury (I/R).^[4] In individuals with SCD, vaso-occlusive crises resulting from hypoxia, I/R injury and subsequent organ damage is common and recurrent. One such frequent presentation, acute chest syndrome (ACS) involving acute lung injury (ALI) is a manifestation of an I/R injury. A large body of literature has been published demonstrating mediation of I/R injuries through unregulated activation of the complement system.^[5, 6] Complementmediated I/R injury commonly occurs when naturally occurring antibodies bind to ischemia neo-antigens initiating complement activation resulting in an avalanche of events generating anaphylatoxins and terminating in MAC formation^[6, 7].

Pharmacological agents to limit antibodyinitiated, complement-mediated pathologic processes are limited. Some commercially available 'complement inhibitors' include the serine protease inhibitor C1 esterase inhibitor (C1-INH) and a humanized C5 monoclonal antibody (Eculizumab). Eculizumab prevents the release of C5a and formation of MAC. It is used for the treatment of paroxysmal nocturnal hemoglobinuria (PNH) and atypical hemolytic uremic syndrome (aHUS). [8] C1-INH is currently only used for treatment of the kinin pathway disease hereditary (familial) angioedema (HAE). The major problem with these drugs is the extremely high cost of producing these very large proteins for medical use. C1-INH also inhibits the fibrinolytic and coagulation pathways, further limiting its utility.^[9, 10] Other complement inhibitors currently in development include the cyclical peptide Compstatin. The parent molecule and its various analogues are in ongoing preclinical and clinical development for application in age related macular degeneration (AMD). Compstatin has been demonstrated to bind to the central molecule C3 inhibiting the classical, lectin and alternative pathway^[11]. Compstatin could potentially increase the risk of life threatening infections due to the profound inhibition of all three complement pathways including the alternative pathway, which is critical for host defenses against pyogenic bacteria.

A novel 15 amino acid complement inhibitory peptide derived from the human astrovirus coat protein is in preclinical development at EVMS/CHKD. This peptide functionally inhibits activation of the C1 molecule ^[12]. Collectively called Peptide Inhibitors of complement C1 (PIC1); subsequent truncations and modifications have yielded the compounds E23A and polar assortant (PA), with improved potency at inhibiting classical pathway activation ^[13].

Preclinical evaluation of PIC1 derivatives included testing of two analogues in an in vitro ABO incompatibility model. The model used normal human serum (NHS) from an individual with blood type O and human red blood cells (hRBCs) from an individual with blood type AB utilizing the presence of

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anti-AB antibodies in the O serum to cause rapid lysis of the AB hRBCs. E23A and PA demonstrated a dose dependent inhibition of hRBC lysis in this model and set the stage for rodent-based in vivo testing in a proof of concept demonstration. [13] Both the analogues of PIC1 crossed the species barrier and inhibited serum complement activation with PA demonstrating complement suppression up to 24 hours post administration. This has provided the impetus for preclinical testing of PIC1 in small animal models of antibody-initiated complement-mediated inflammatory diseases including AIHTR^[14], ALI in SCD, and hypoxic ischemic encephalopathy (HIE) ^[15]. Complement-inhibitory interventions, like PIC1, have significant therapeutic potential to limit inflammationmediated end organ damage in patients with SCD and will be a primary focus of future investigations.

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Ethical, Legal and Financial Implications of Providing Interpretation Services for Patients and Parents with Limited English Proficiency

Objective: Participants will be able to describe the ethical framework for the provision of interpreter services to LEP potential families. List major pieces of legislation and regulatory bodies which governs the use of interpretation in medical practice. Analyze the financial implications of providing or not providing interpreter service to their patients.

ACGME Competencies: Patient Care, Interpersonal Communication Skills, Professionalism, Systems-based Practice



Introduction In 2012, nearly 25 million Americans age 5 and over were estimated by the U.S. Census Bureau to have limited English proficiency (LEP) (with limited English

proficiency defined as speaking no English or speaking it less than "very well"). In fact, the same survey indicates that only 79% of US households speak English exclusively.ⁱ In Hampton Roads, the number of individuals age 5 and over with LEP is estimated to be around 40,000. In the 5-17 year-old age range alone, the Census Bureau estimates that there are almost 21,500 children in Hampton Roads whose primary language is not English." The majority of these children are Spanish-speaking. In reality, these numbers are likely even higher since many undocumented immigrants choose not to participate in surveys for fear of deportation or other forms of retaliation.

Ethical Implications

Even when providing treatment to native English speakers, there is near-universal agreement that accurate communication decreases the occurrence of medical errors. When the patient/parent and provider do not speak the same language, the potential for medical errors increases. Based on their review of the literature, Wasserman et al. concluded that "LEP patients have been harmed by poor comprehension of their medical condition, treatment plan, discharge instructions, complications, and followup; inaccurate and incomplete medical history; ineffective or improper use of medications or serious medication errors; improper preparation for tests and procedures; and poor or inadequate informed consent" (p. 5).ⁱⁱⁱ Stories like that of Kamil Čák, DMin, BCC Director, Chaplaincy, Bioethics, Cultural Language Services, Patient Relations Children's Hospital of The King's Daughters Assistant Professor of Pediatrics Eastern Virainia Medical School

13-year-old Graciela Zamora have now been corroborated by research which has demonstrated, for example, that Latino children living in LEP households have significantly higher rates of appendiceal perforation than children living in English-speaking households.^{iv} Graciela died from sepsis caused by a ruptured appendix because her parents did not understand the discharge instructions which were given to them without an interpreter.^v If medicine's goal is primum non nocere, then the logical conclusion is that the only ethically appropriate choice is to provide adequate interpretation services for LEP patients and their parents. The remaining 3 of the 4 basic bioethical principles (autonomy, beneficence and justice) would also unequivocally support this conclusion. After all, a parent or patient are unable to make an autonomous decision without understanding the illness and its consequences; providing an interpreter is the beneficent choice and it is practically impossible to deliver care justly if everyone does not have equal access to care and information.

Legal Implications

Providing qualified medical interpreters for LEP patients/parents is also the legally appropriate course. A number of federal and state laws, as well as regulatory requirements, address the patients' rights and healthcare providers' responsibilities to provide such services. All in all, there are more than 10 Virginia laws that address language access in health care facilities. The basis for all legal requirements is Title VI of the 1964 Civil Rights Act which prohibits discrimination based on race, color or national origin.^v Both federal agencies and the Supreme Court have consistently interpreted language needs as falling under the umbrella of national origin.v The second key piece of legislation is the Americans with Disabilities

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Act (ADA) which specifically applies to the deaf and hard of hearing patients/families. Title III of the ADA, "prohibits discrimination on the basis of disability in the activities of ... doctors' offices."^{vii} The law does not absolve any healthcare providers from fulfilling this responsibility based on the size of the office, practice or facility and specifies some situations when an interpreter is required:

• discussing a patient's symptoms and medical condition, medications, and medical history

 explaining and describing medical conditions, tests, treatment options, medications, surgery and other procedures

• providing a diagnosis, prognosis, and recommendation for treatment

obtaining informed consent for treatment

• communicating with a patient during treatment, testing procedures, and during physician's rounds

• providing instructions for medications, post-treatment activities, and follow-up treatment

• providing mental health services, including group or individual therapy, or counseling for patients and family members

• providing information about blood or organ donations

• explaining living wills and powers of attorney

discussing complex billing or insurance matters

 making educational presentations, such as birthing and new parent classes, nutrition and weight management counseling, and CPR and first aid training viii

One of the most clearly and directly written standards for language access in healthcare was published by the U. S. Department of Health and Human Services (HHS) Office of Minority Health and states that providers are to: "offer language assistance to individuals who have limited English proficiency and/or other communication needs, at no cost to them, to facilitate timely access to all health care and services."^{ix}

Financial Implications

While providing foreign language interpretation is the ethically and legally appropriate healthcare practice, this practice clearly comes with associated costs, not the least of which is financial. Providers are specifically prohibited from passing interpretation costs

on to their patients. And while "in 2000, the Centers for Medicare & Medicaid Services (CMS) reminded states that they could include language services as an administrative or optional covered service in their Medicaid and State Children's Health Insurance Programs, and thus directly reimburse providers for the costs of these services for program enrollees,"x Virginia only reimburses interpreter charges incurred at a few state-run clinics and North Carolina does not participate in the reimbursement program at all. Moreover, CMS makes it clear that any provider receiving public funds (i.e., Medicare or Medicaid dollars) is required to provide foreign language interpretation to all patients and families served by that provider. The ADA does allow for an exception from the provider's requirement to furnish interpretation services for the deaf or hard of hearing. However, this exception is mostly symbolic in nature. The National Association of the Deaf (NAD) states, "The ADA does not require the provision of any auxiliary aid or service that would result in an undue burden or in a fundamental alteration in the nature of the goods or services provided by a health care provider." However, the NAD quickly adds the following statements: "Making information or communication accessible to an individual who is deaf or hard of hearing is unlikely ever to be a fundamental alteration of a health care service," and "showing an undue burden may be difficult for most health care providers."xi

Of course, choosing not to utilize interpreters can be costly as well. Some of the better-known adjudicated cases of LEP patients who were harmed as a result of misunderstandings include "the misdiagnosis of a cerebral haemorrhage in a patient that resulted in a \$71 million award for damages" or the 1990 U.S. case of "a Mexican labourer . . . [who] reportedly lost sight as a result of a work-related eye injury [which was improperly] addressed, diagnosed, and treated by an attending doctor at a clinic."xii The patient was awarded \$350,000.

Finally, when families with LEP do not have access to interpreters to help them apply for services such as emergency Medicaid, providers are left without payment for services that could have been covered by a governmental insurance program.

Recommendations

1. Healthcare providers (regardless of the size of their practice) must be aware of and meet their obligation to provide language assistance to their patients/families with limited English proficiency.

2. Healthcare providers should contract with qualified (certified) interpreters to provide either in-person, over-the-phone or video remote interpretation.

3. Smaller practices who may need guidance in establishing language assistance services should contact their state Health Department or the Language Services Department of a local health system for guidance.

4. When using a qualified interpreter to address a patient's language need, the provider should always note this in the medical record and include the interpreter's name or number in the note.

5. Healthcare providers should engage in advocating for state and federal reimbursement for the costs associated with providing medical interpretation.

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Galactosemia and the Newborn Screen

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Objective: Participants will be able to review the physiology of Galactosemia, as well as the clinical presentation of critical newborns with the disorder and list the necessary steps after receiving a critical or abnormal scores for Galactosemia.

ACGME Competencies: Patient Care, Medical Knowledge and Practice-based Learning and Improvement

Introduction

This year marks the 50th anniversary of the advent of newborn screening in the United States, an endeavor that has revolutionized the recognition and treatment of potentially lethal disorders in the neonatal period. The number of disorders screened varies from state to state, but a minimum core panel of 31 genetic, endocrine, and hematologic diseases are tested in the first two days of life. More diseases will likely be added as diagnostic technology advances.

Fortunately, the general pediatrician is unlikely to encounter many true positives each year. However, one of the most frequent abnormal or critical results that a primary care physician may see is for galactosemia. Here, we will briefly review this disorder, the laboratory tests used to evaluate the disease, and what the pediatrician's role is in caring for these children.



What is Galactosemia?

Galactosemia is an inborn error of metabolism involving the breakdown of the sugar galactose in the body. Galactose and glucose form the disaccharide lac-

tose, which is the main carbohydrate in milk. There are currently three known inborn errors of galactose metabolism, the most serious being classic galactosemia, caused by deficiency in the galactose-1-phosphate uridyltransferase (GALT) enzyme. Each of the three disorders in the galactose pathway result in the inability to process lactose in breast milk or cow's milk formula. If galactose cannot be fully metabolized, elevations

of both galactose and toxic by-products in the pathway cause damage. Complete or near-complete GALT enzyme deficiency can lead to severe morbidity and mortality if left untreated. Symptoms may occur in the first or second week of life after ingestion of breast milk or formula. Infants may present with failure to thrive, vomiting, jaundice, lethargy, and refusal to feed. Hepatomegaly, ascites, liver cirrhosis, and death from sepsis (usually from E. coli) may follow. Cataracts may also appear within days or weeks. Longterm complications include increased risks for developmental or cognitive delay and difficulties with speech. Females with classic galactosemia often have infertility due to premature ovarian failure.

Classic galactosemia has a prevalence of one in 40,000-60,000 in North America, and is inherited in an autosomal recessive pattern. Over 167 different mutations have been reported in the GALT gene located on chromosome 9. Genotype-phenotype correlations are available, with some mutations causing a more severe presentation.

The other two types of the disease include galactose epimerase (GALE) deficiency and galactokinase (GALK) deficiency. GALE deficiency may have a similar presentation to GALT deficiency, but is considered extremely rare. The only consistent feature of GALK deficiency is cataracts; damage to the brain, liver, and kidneys is not typically seen. It is also less common than GALT deficiency, occurring in about one in 150,000 to 1 million. Both GALE and GALK deficiencies may be more common in specific isolated populations or in populations with higher frequency of consanguinity.

How is Galactosemia screened?

There are two markers for galactosemia on the Virginia newborn screening panel. The

Beutler screen measures galactose-1-phosphate uridyl transferase (GALT) enzyme activity by quantitative fluorometric testing. The Hill screen measures the galactose-1-phosphate level. The Beutler screen is very sensitive to environmental conditions such as heat and humidity and the presence of these conditions may result in false positive results.

What is the primary physician's role?

The approach to follow-up on an abnormal galactosemia screen is dependent on the threshold of results. In the Commonwealth of Virginia there are two levels of concern that prompt a requirement for follow-up. The first type of result is termed "abnormal." An abnormal result only requires the primary physician to repeat the newborn screening card. The second type of result is termed "critical." "Critical" levels signify a higher likelihood that an infant has a true metabolic disorder and needs immediate medical attention. In this situation the VDH newborn screening nurse will recommend that the metabolic geneticist be consulted, and they will request your permission to place that consultation directly. The geneticists' office will then recommend follow-up testing to the primary physician. In order for an infant to have a critical result for galactosemia they must either have 3 consecutive abnormal Beutler levels or both an abnormal Beutler and an abnormal/critical Hill level on the same screen.

An infant with a critical newborn screen for galactosemia should be seen in the pediatrician's office within 24 hours of being notified of the screen results. The infant should be evaluated for feeding, growth, and general well-being. If there are any concerns that the infant is showing signs of classic galactosemia or illness the metabolic geneticist should be contacted immediately. If the

continued from page 14...

infant appears healthy they can be sent for follow-up laboratory studies and then switched to soy formula. Typically an appointment in the metabolic clinic is not scheduled until after the follow-up lab results are available (on average 2-3 weeks).

In general, we do not recommend switching a child to soy formula unless they have had a critical screen for galactosemia where the Hill level is elevated.

The follow-up laboratory studies need to be completed prior to starting soy formula so that a baseline pre-treatment galactose-1-phosphate level can be obtained.

Follow-up diagnostic studies are generally not recommended unless the infant has either a critical screen for galactosemia or hashad two consecutive abnormal screens. Diagnostic laboratory studies should include: GALT enzyme activity and phenotyping, galactose-1-phosphate level, urine galactitol level, and a Perkin-Elmer comprehensive newborn screen with DNA screen for galactosemia. It is crucial that this entire set of labs is obtained because they are necessary for the accurate differentiation between classic galactosemia, milder variants such as Duarte galactosemia, and carrier states.

Metabolic Clinics in Virginia There are three metabolic centers in the Commonwealth who receive newborn screen results for each of their encatchment 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: Arti Pandya, 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



HPV Vaccine is Cancer Prevention

(and may eliminate the next generation of children with recurrent respiratory papilloma disease)

Craig Derkay, MD, FAAP, FACS

Professor and Vice-Chairman Department of Otolaryngology and Pediatrics Eastern Virginia Medical School Director, Pediatric Otolaryngology Children's Hospital of the King's Daughters Norfolk, Virginia

Human Papilloma Virus (HPV) infection is widespread in the United States with more than 79 million Americans infected with the virus and more than 14 million new infections annually, according to the Center for Disease Control (CDC). About three-fourths of these infections will occur in 15-24 year old children and young adults usually within 4 years of the first sexual intercourse. HPV is responsible for causing 4000 deaths from cervical cancer yearly and more than 26,000 HPV-related cancers (8500 in males and 17500 in females) annually. Many people who have HPV do not know it because the viral infection often has no significant signs or symptoms and is passed to partners without knowing it. There are more than 100 closely related sub-types of HPV with HPV 16 and 18 most commonly associated with cervical, penile, anal and head and neck cancers and HPV 6 and 11 most commonly associated with development of genital warts and recurrent respiratory papillomas (RRP).

As a Pediatric Otolaryngologist, we are confronted with children who have been exposed to HPV as they traverse the birth canal and present to us typically before their 5th birthday with hoarseness, progressive hoarseness and varying degrees of respiratory distress due to RRP (photo). There is typically a lag of about a year between the initial onset of symptoms and definitive diagnosis with a number of these children labeled as "asthmatic" or "recurrent croup". The prognosis for these children is very variable with some requiring only a few lifetime surgical procedures while others may need more than a hundred. There is currently no adjuvant medical management that has proven successful in managing RRP while

Objective: Participants will be able to discuss the importance of offering HPV Vaccinations to their adolescent boys and girls. **ACGME Competencies:** Patient Care, Professionalism, Medical Knowledge, and Interpersonal Communications Skills.

10-15% will develop disease in their trachea; 1-2% in their lungs and mortality of 1-2% due to either airway obstruction, malignant degeneration, chronic lung disease or an anesthesia disaster.

In 2006, the FDA approved a guadrivalent (HPV 6,11, 16, 18) HPV recombinant vaccine (Gardasil, Merck) for girls between 9-26 years of age. This vaccine was found to be highly effective in preventing cervical cancer, cervical pre-cancers, cervical dysplasia and genital warts. In 2011, the ACIP extended the recommendation for vaccination to include boys 9-21 years from permissive to routine to prevent genital warts, penile and anal carcinomas. Since approval, more than 56 million doses have been administered in the U.S. however, only 50% of the eligible girls and far fewer boys are currently being vaccinated. This is in spite of close safety monitoring of the vaccine demonstrating an excellent safety profile and economic modeling that has demonstrated that every 13 cents spent on vaccination results in a reduction of \$1 in health care costs for treatment of HPV related diseases. Early evidence of vaccine effectiveness is emerging from Australia, Denmark, and Sweden where school-based vaccination programs and universal health care programs are providing herd immunity with a measurable reduction in genital warts and cervical precursor lesions. In the words of Larry Pickering, MD, Chair of the ACIP, "Don't let your pediatric patients become an Oncologist's patient in 20 years!"

Unfortunately, the current vaccine is not effective at clearing existing infections so it is imperative to administer the vaccine before children become sexually active. Many studies have shown that there is no effect on sexual mores as a result of discussion and counseling about the benefits of administering this vaccine (much like utilizing seat belts does not increase the incidence of drunk driving).

From an Otolaryngologist's standpoint, we are very hopeful that achieving vaccination uptake at the level of 75-80% will result in a sufficient level of herd immunity to wipe out the next generation of RRP patients much like the introduction of the HiB vaccine in the late 1980's virtually eliminated acute epiglottitis in our Children's hospitals.

The AAP has received a grant from CDC to create webinars, online quality improvement modules and other resources for Pediatricians on how to improve adolescent vaccination rates for HPV. Please present the vaccine to the families in your practices with the conviction and urgency that it deserves and not just suggest or mention it—families are looking to their Pediatricians for guidance in this area.



PEDS 101

John W. Harrington MD Division Director of General Academic Pediatrics Children's Hospital of The King's Daughters

Objective: Participants will be able to discuss how to improve resident awareness along with comfort and knowledge base in areas that increase infant mortality. **ACGME Competencies:** Patient Care, Practice-based Learning and Improvement, Professionalism, Medical Knowledge, Interpersonal Communications Skills and System-based Practice.

What started as a pet project from Dr. Donald Lewis and Dr. Karen Remley has now been a sustainable annual event that is produced for our incoming Pediatric Interns. We have called it Pediatrics 101 or PEDS 101. Essentially it is a two week boot camp around some of the most common pediatric problems and common advocacy areas that young pediatricians need expertise early on in their careers. The topics include breastfeeding, safe sleep/sleep hygiene, vaccines, over the counter medications, complementary therapies, car seat safety, child-proofing, developmental and mental health screening, recognition of child abuse, nutrition after breastfeeding, and common behavioral strategies.

These areas are covered with some didactic lecture, hands-on training, and the use of role play and standardized patients. The two week period is broken up into eighteen one-hour slots, with an 8am and a noon session Monday through Friday and Grand Rounds on Thursday mornings. Most of the concepts are separated into a didactic morning session with a practical part in the afternoon. For example, the breastfeeding dyad has a lecture in the morning and in the afternoon we have developed two stations: Station 1 provides the residents and medical students a chance to view and understand the breastfeeding paraphernalia of pumps, nipple shields, and how to do cross-cradle versus football holds. Station 2 provides a standardized patient scenario that depicts a new young mother with an infant that she would like to breastfeed, but her grandmother is preventing the process and the resident must intervene. The vaccine dyad is also similar with a didactic lecture in the morning and then two stations set up in the afternoon. Station 1 provides the residents the chance to actually give vaccines and place PPDs into food products such as chicken legs and hotdogs/sausages. Station 2 provides a standardized patient scenario where hot parents have decided that they would rather not immunize their 2 month old because they think it may be harmful and the resident must convince them of the benefits of vaccines.

These strategies provide our incoming residents with the real life experiences and some skill sets that they will need when they are first introduced to the outpatient clinics. Having a chance to approach these topics in a supportive environment and be given feedback early on has been extremely important to our residents. Learning tactics related to controlling the room or motivational interviewing are just as important as starting an IV or taking a history. This two week course has now also become part of our Master in Public Health certificate curriculum, which is currently one of only six programs in the country. The residents describe the two week period as fun and extremely useful so early in their clinical careers and statistically speaking it appears to be improving their comfort and knowledge on the topics covered.

For those pediatricians who practice in an academic environment with trainees (including residents, medical students, Nurse Practitioner Students, Physician Assistant students), a similarly structured program may enhance the trainees experience as well as their effectiveness with patients. For those in other settings, a shortened structure may help acclimate new nurses, medical assistants, or other office staff to the pediatric outpatient environment.



The Dangers of Synthetic Cannabinoids

Sanaz B. Devlin, MD Pediatric Hospitalist Children's Hospital of The King's Daughters

John J. Devlin, MD Emergency Physician/Medical Toxicologist Emory University School of Medicine

Myocardial infarction, renal failure, seizures - these significantly morbid conditions have all been associated with synthetic cannabinoid use in previously healthy adolescent patients.^{1,2,3} Teenagers are frequently duped into believing these chemicals are healthy and/or legal alternatives to marijuana use due to their availability and relative ease of purchase. Synthetic cannabinoids are sold in convenience stores and head shops as incense, air fresheners, or potpourri under a variety of names such as Spice and K2, typically with the label "not for human consumption." This "synthetic marijuana" actually consist of laboratory-engineered chemicals sprayed onto vegetable matter by drug dealers and sold to teenagers to be smoked like marijuana cigarettes. Poisoning can result from inherent toxicity of the synthetic cannabinoids, the vegetable material, or products of pyrolysis.¹ Many synthetic cannabinoids became schedule I in 2012 under the Synthetic Drug Abuse Prevention Act. The synthetic cannabinoids JWH-018, JWH-073, and AM-2201 are the most commonly identified but novel chemicals are introduced into the illicit drug market as quickly as the older agents are scheduled by the Drug Enforcement Agency.

Cannabis is widely used worldwide and in the United States. Therefore, it is not surprising that nationwide 40.7% of adolescents in 9-12 grade have used cannabis one or more times in their lifetime.⁴ A survey of marijuana users reveals that half have also smoked synthetic cannabinoids and almost one-quarter report current use.⁵

Synthetic cannabinoids mimic the effects of Δ^9 -tetrahydrocannabinol (THC), the active compound in cannabis, but are 5-800 times more potent as agonists of the central nervous system's cannabinoid receptors.^{6,7} Compared to cannabis, the adverse effects of synthetic cannabinoids are more severe and more diverse. Because of this chemical structure heterogeneity and lot-to-lot concentration variability, the clinical effects of synthetic cannabinoids are a spectrum, varying from mild to severe. CNS effects are prominent and include seizures, agitations, anxiety, sedation, and confusion.⁷ The psychological effects of synthetic cannabinoids, such as psychosis, appear to be more severe than with cannabis. Recently, acute kidney injury resulting in hemodialysis has been associated with smoking synthetic cannabinoids.¹ Cardiovascular effects are common, typically tachycardia and hypertension consistent with a sympathomimetic toxidrome. However, there have been cases of myocardial infarction in otherwise healthy teenagers after use of synthetic cannabinoids. In these three cases, inferior or inferolateral ST segment elevation was identified on electrocardiography in the context of serum troponin elevation.3 Other reported findings include nausea, vomiting, mydriasis, conjunctivitis, and rhabdomyolysis.⁷ There is also an increased incidence of addiction to synthetic cannabinoids. Symptoms of withdrawal include nervousness. irritability. sleeping difficulties, strange dreams, and diaphoresis.

Synthetic cannabinoids are not detected on routine urine drug screens.⁷ However, a positive urine drug screen for THC in the context of a bizarre presentation after "marijuana" use should prompt the clinician to consider synthetic cannabinoid exposure given the common practice co-abuse of cannabis and synthetic cannabinoids. Confirmatory tests for suspected exposures to common synthetic cannabinoids are available at specialized reference laboratories. Other findings on routine chemistries that have been associated with synthetic cannabinoids include hypokalemia and hyperglycemia.⁸



Objective: Participants wil be able to discuss clinical manifestations of synthetic cannabinoid use. Discuss evaluation and management of patients who have been exposed to synthetic cannabinoids.

ACGME Competencies: Patient Care, Medical Knowledge

Management guidance for synthetic cannabinoid toxicity is available via telephone from all poison control centers (1-800-222-1222). Management is supportive as no antidote exists.⁷ Sympathomimetic effects, such as tachycardia, hypertension, and agitation, can be treated with benzodiazepines, such as diazepam, lorazepam, and midazolam. Additionally, synthetic cannabinoid-induced seizures typically respond to benzodiazepines and status epilepticus is uncommon. Weight-based intravenous fluids may be helpful for rhabdomyolysis and dehydration from vomiting.

The synthetic cannabinoid problem won't be resolved quickly. National poison center data indicates that usage is increasing. Adolescents will continue to be reluctant to share their drug use history and may not even realize that these synthetic chemicals are responsible for their medical conditions. Therefore, it is imperative that pediatricians become familiar with these products and are comfortable screening their patients for use.

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Resource Mothers (Madres Tutalares) -

A Model Program to Improve Prenatal Care and Birth Dutcomes in a Barrio in Santo Domingo, Dominican Republic

Leslie Toledo, MPH Program Director, Physicians For Peace Norfolk, Virginia Korinne Chiu, PhD Director of Program Evaluation and Organizational Leaning, Physicians for Peace Edward H. Karotkin, MD, FAAP Professor of Pediatrics The Eastern Virginia Medical School

Objective: Participants will be able to describe the Resources Mother's Program (Madres Tutalares) a model program to improve prenatal care and birth outcomes in a Barrio in Santo Domingo, Dominican Republic. **ACGME Competencies:** Patient Care, Medical Knowledge

Every year in developing countries, 7.3 million girls under the age of 18 give birth (UN-FPA State of the World Population 2013). To address this public health concern, a socio-ecological framework addressing the needs of adolescents within their communities is ideal to promote health, education, and productivity. In 2005, a U.S.-based nonprofit, Physicians for Peace (PFP), partnered with a local non-profit in Santo Domingo, Dominican Republic, Fundación Sol Naciente (FUSNI), to implement a program for pregnant and parenting teenagers living in nearby barrios using a community-based participatory approach modeled after a U.S.based program.

The program implemented in the Dominican Republic is modeled after the Virginia-based Resource Mothers program, instituted at The Children's Hospital of The King's Daughters and The Eastern Virginia Medical School, which was developed in 1998 to improve neonatal and perinatal health of African American women. In South Hampton Roads, indicators of maternal and neonatal health are worse than in other regions of the state and country and African American neonatal and infant mortality rates, and low birth weight rates are higher than the white population. During the pilot program, outcome data, including birth weights, neonatal mortality rates, infant mortality rates, and lengths of hospital stay, were collected and compared to controls. Over a two and a half year period, 589 women were served and 284 infants were delivered. During that time, the incidence of very low birth weight (<1500gm) was lower in the Resource Mothers group, 2/1000 live births vs. 4/1000 live

births in the controls. The incidence of extremely low birth weight infants (<1000gm) was lower in the Resource Mothers group, 0.5/1000 live births vs. 1.5/1000 live births in the controls. There were no neonatal deaths (deaths prior to 28 days of age) in the resource Mothers group vs. 13.1/1000 live births in the controls. Infant mortality rates, in the Resource Mothers group were lower, 10.1 1000 live births vs. 14.8/ 1000 live births in the controls.

Translating this successful US-initiated program to the resource-poor barrios of Santo Domingo, and targeting services to teens, involved modifications in curriculum, innovations in data collection and reporting, propelling of local health champions and adaptation of social safety nets. These adaptations were identified and implemented using an external evaluation and thorough needs assessment to better understand need and local resources. The number of people living in Santo Domingo's barrios, makeshift communities lacking basic infrastructure such as clean water, sanitation, is on the rise. Santo Domingo has the greatest migrant population in the country, adding to the overwhelmed health care system that struggles to stretch limited resources to meet the population's need for quality health services. Particularly vulnerable are pregnant adolescents, who are at higher potential risks for several clinical complications compared to adult women. According to the 2013 UN Report Caribbean 360, News around the Caribbean, an estimated 70,000 adolescents in developing countries die each year from complications during pregnancy and childbirth. The

risks associated with adolescent pregnancy include low birth weight, poor weight gain during pregnancy, premature birth, perinatal death, cesarean delivery, preeclampsia, irondeficiency anemia, lack of social support and low income. The PFP Resource Mothers program addresses these health risks by ensuring weekly home visits and accompanying young mothers to their prenatal appointments. 2013 outcomes show that out of 119 teen mothers, 90% attended at least six prenatal appointments, premature births occurred at a rate of less than 8% and 75% of teens in the program report having breastfed.

The PFP Resource Mothers program pairs experienced mothers with pregnant teens, who, in their role as the teen's mentor, provide education, emotional support and accompaniment to prenatal and well child medical appointments during the teen's pregnancy and throughout the first year of life. Resource Mothers (RMs) are appointed by the community using established criteria and because they live in the barrios where they work, RMs are able to identify and recruit teens early in their pregnancy.

RMs conduct weekly home visits teaching pregnant teens on prenatal and newborn health topics as well as guidance and emotional support to navigate the healthcare system. Mentor training is provided by health professionals at the local maternity hospital, endorsing the Resource Mothers program as a vital maternal outreach activity of the health system. Ongoing training and health education is provided by PFP and local health partners.

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The alignment of PFP with FUSNI helps to further the relationships between the local program and the Maternity Hospital of San Lorenzo de Los Mina, which remains the primary hospital location where the RM clients attend prenatal visits and deliver their babies. The RM program maintains an office at FUSNI where program data, meetings and local training take place. The current FUSNI location is close in proximity to the Maternity Hospital, aiding the ability of the RMs to successfully manage their responsibilities of counseling and education, accompanying clients to prenatal and infant medical appointments as well as collecting and delivering program data.

The PFP Resource Mothers program began with 10 Resource Mothers (RMs) working in 5 barrios in Santo Domingo, working with 92 pregnant teens in 2005. In 2008, the program expanded to 20 RMs in 10 barrios. Today, 19 RMs serve approximately 150-250 pregnant and recently delivered teenagers each year. Four of the RMs serving today are past clients, who offer the unique perspective of client and mentor, which exemplifies the role of empowerment and community leadership that the program upholds. The trusting relationship between RMs and teen clients enhances program compliance and aids in the delivery of prenatal and postpartum education conducted during weekly home visits. To date, the program has served over 670 pregnant teens.

In order to assess program progress, measures were piloted and validated with the RMs to collect data at four different time points throughout the program. These

participatory monitoring and evaluation measures assess program process indicators and maternal and child health outcomes against agreed-upon program targets. Data are entered locally into an online database and analyzed quarterly to assess progress toward program objectives. Quantitative and qualitative data are analyzed and interpreted with local partners in order to assess current program needs, adapt program areas to the local context, and identify program successes. Based on revised data collection efforts, in the fourth quarter of 2013, 75% of teens in the program reported having breastfed and premature births occurred at a rate of less than 8%. Use of family planning methods nearly doubled from 43% to 84% by graduation from the program. At year-end 2013, over 50% of teens in the program reported continued breastfeeding at 12 months post-partum. Systematic monitoring of indicators and outcomes enables program staff to make revisions or improvements to the program based on up-to-date program information.

Summary:

The Resource Mothers program is a low tech, low cost intervention that improves the outcome of pregnancy by increasing access to prenatal care, decreasing low birth weight, lowering neonatal and infant mortality and improving many of the common indicators of pediatric health. Implementing evidence-based strategies and adapting to local needs has evolved to incorporate unique program strengths such as on-going training and education, participatory monitoring and evaluation and local champions to increase health benefits. The mentor relationship and emotional bond between RM and teen mother are examples of how hightouch, low tech solutions enhance program quality. This program has the potential for serving as a model for other communities committed to improving the outcome of pregnancy.

Leslie Toledo, MPH, Program Director, Physicians for Peace Korinne Chiu, PHD, Director of Program Evaluation and Organizational Learning, Physicians for Peace

VA-AAP NEWSLETTER REGISTRATION AND EVALUATION FORM (Summer 2014)

You have the opportunity to claim up to 3.0 AMA PRA Category 1 Credit(s) $^{\text{m}}$.

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This CME activity will expire on September 30, 2015.

Please send form to: Rosalind Jenkins, c/o CHKD, 601 Children's Lane, Norfolk, VA 23507 Please allow up to 8 weeks to receive your certificate.

Hospital-based Community Garden Project

With the support of The Children's Hospital of the King's Daughters (CHKD) and Eastern Virginia Medical School (EVMS), first-year Pediatric Residents have set up a hospital-based community garden project. The intent of this novel endeavor is to improve families' attitudes towards healthy eating with a series of hands-on classes and activities. Twelve participating children between the ages of 8 and 12 from CHKD clinics located in the Norfolk community, as well as their parents, are learning what it takes to create a sustainable food source in a fun and educational way. This IRB-approved project takes advantage of the multiple resources available at CHKD, including experienced faculty, registered dietitians, and enthusiastic medical students all under the leadership of first year pediatric residents. Over a three-month period the participants will be taking classes during which they will learn about the plants they are growing and why a healthy diet is important to all aspects of their lives. Classes cover a variety of topics all with the goal of helping families and children make simple, yet healthy choices at mealtime. Example of the weekly classes include advice on shopping for and cooking a healthy meal on a budget, exposing the families to a variety of fruits and vegetables (many of which they have never tasted before), and the basic biology of plant development; all while returning to the central theme of creating a garden-to-table meal. This pilot project has received great support from the community and the CHKD faculty and administration. While the project is still in its first season, the response from the participants and the feedback from other community healthcare providers has been nothing short of excellent. Therefore the project organizers look forward to the opportunity to invite Tidewater children and their families to participate in future CHKD community garden endeavors for years to come. This project is a perfect example of the combined missions of CHKD and EVMS by blending resident leadership with community involvement to establish a medical home that will be staffed by the pediatricians of tomorrow.

Peter Farrell MD, Michael Rogers DO, Kyle Brady DO



D.W. Lewis Memorial Golf

Tournament Presented by the Department of Pediatrics

Saturday Sept 13th 1 PM Start, Noon Check In Bide-a-Wee Golf Course Portsmouth, VA

\$125 per player (\$75 per resident/\$50 per student)
Casual dinner to follow
Four player traditional scramble
Prizes
Hole sponsorships (\$100)
Long drive/closest to pin
sponsorships (\$100)

All proceeds benefit the Chairman's Fund for Ha



Register on attached form or via email by Sept 5th <u>LewisMemorial@chkd.org</u>

For more info, visit http://pediatrics.evms.edu/residency/golf.html

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