



VIRGINIA • PEDIATRICS

American Academy of Pediatrics • Virginia Chapter

IN THIS Issue WINTER 2019

- 2 PRESIDENT'S MESSAGE
- 3 CME INFORMATON
- 4 PTSD IN NICU MOTHERS
- 5 NEONATAL MEDICINE: THREE QUALITY CARE IMPROVEMENT STRATEGIES TO INFLUENCE HEALTH OUTCOME OF VA'S MOST VULNERABLE INFANTS
- 8 ESSENTIALS OF PEDIATRIC BURN INJURY
- 11 QUIET PLEASE! QUALITY IMPROVEMENT PROJECT TO MINIMIZE SLEEP DISRUPTIONS IN STABLE INPATIENT PEDIATRIC POPULATION
- 13 PEDIATRIC ISOLATED LINEAR SKULL FRACTURES:
- 14 RED BLOOD CELL TRANSFUSION STRATEGIES IN CHILDREN
- 16 REVISED GUIDELINES: METABOLIC / BARIATRIC SURGERY IN YOUTH WITH SEVERE OBESITY
- 17 STANDARDIZED APPROACH TO CF CARE AT CHOR-VCU
- 22 SCREENING FOR SOCIAL DETERMINANTS
- 23 PRIMER ON TICKBORNE RICKET
- 24 CME REGISTRATION AND EVALUATION FORM
- 25 IMPROVING ACCESS TO MENTAL HEALTH SERVICES
- 26 PLAY: A CRITICAL SKILL FOR INFANTS/CHILDREN
- 27 NEW CHOR GUIDELINES ACCESSIBLE TO ALL!
- 27 STUDYING MISSED WELL CHILD CARE ACROSS USA
- 28 THE NEED FOR MULTI-DISCIPLINARY CLINICS IN PEDIATRIC SUB-SPECIALTIES
- 29 IMPROVING CARE FOR SUBSTANCE EXPOSED INFANTS
- 29 PEDIATRICS TRAUMATIC BRAIN INJURY PROGRAM
- 30 PEDIATRIC GENERAL ASSEMBLY DAY REGISTRATION

About Us AAP-VIRGINIA Chapter

We welcome your opinions and ideas.

Please send comments on articles, ideas for new articles, letters to the editor, suggestions for making Virginia Pediatrics more useful and address changes to:

Virginia Pediatrics

Jane Chappell: Executive Director
2821 Emerywood Pkwy | Suite 200
Richmond, VA 23294

- P (804) 622-8135 • F (804) 788-9987 •
- email: jchappell@ramdocs.org •
- www.virginiapediatrics.org •

- Next Issue: Spring 2019 •
- Deadline for entries: 3/5/2018 •

Publication of an advertisement in Virginia Pediatrics neither constitutes nor implies a guarantee or endorsement by Virginia Pediatrics or the VA-AAP/VPS of the product or service advertised or of the claims made for the product or service by the advertiser.

:: newsletter design services : Flutterby Media ::
Royalty Free Images: Storyblocks where not otherwise noted.

Firearm Safety: How is Your Community Getting Involved?

2018-2019 Leonard P. Rome CATCH Grant Recipients

Rebecca Furby, MD, Adrienne Kirby, MD, Jennifer Chmielewski, MS4

Today, our children and communities are becoming more affected by gun violence. With tragedies such as the Sandy Hook Elementary massacre and the Parkland School shooting, we must recognize that gun violence is affecting children of every age nationwide. Gun violence has hit home for us as well. In 2016, 78 Virginians, aged 0-19, died as a result of firearms, translating to 4,603 years of potential life lost. Eastern Virginia was consistently the region with the highest number of firearm-related deaths among 0-19 year olds from 1999-2016.

What can we, as pediatricians, do to prevent such tragedies? And where do you start?

It takes a village to raise a child. In efforts to improve the safety of our children, the Children's Hospital of the King's Daughters' Resident Advocacy group wanted to involve the community. By doing so, we hoped to address community concerns for youth safety, develop innovative solutions, receive buy-in from key community leaders, and hopefully raise enough voices to change legislation. But how do you unify a hospital, local schools, and a community under the same vision? We hoped to learn more about this process from our Visiting Professor, Dr. James Wright.

As the founding director of the Child Health Advocacy Institute (CHAI) in 2007 at Children's National Health System, he has been at the forefront of community and hospital collaborations. With passion and dedication for addressing pediatric disparities, CHAI is now a pediatric advocacy center of DC with ongoing community outreach projects within schools, educative programs for parents and patients alike, consultation services for local pediatricians, and pediatric resident advocacy initiatives. With such impressive accomplishments as well as expertise as a Pediatric Emergency Medicine physician, chair for the AAP Committee on Pediatric EM, and inaugural chair of the AAP Violence Prevention Subcommittee, Dr. Wright was invited to help guide our discussion and efforts in preventing pediatric gun violence. We knew his expertise could assist us in further developing the cherished building blocks of the Children's Hospital of the King's Daughters to become a pediatric advocacy center for Hampton Roads.

In our two-day Visiting Professorship, Dr. Wright spoke with key leaders about the development of an advocacy curriculum and program

management within the health system. He additionally provided a stepwise approach to hospital integration and community involvement surrounding advocacy efforts.

1 Data from Virginia Department of Health via Virginia Online Injury Report System: <http://www.vdh.virginia.gov/voirs/injury-deaths/>

As CHKD is a newly certified Level 1 Pediatric Trauma Center, Dr. Wright also led discussions on pediatric trauma. A highlight during Dr. Wright's time at CHKD was a panel discussion led by community members dedicated to firearm safety. Our panel members included a pediatric trauma surgeon, a physician and Chair of the Norfolk School Board, a community reverend, and the legislative lead for the Hampton Roads division of Moms Demand Action (MDA).

During the panel discussion, we were introduced to the BeSmart campaign, which is promoted by Moms Demand Action. MDA is a group dedicated to implementing public safety measures that will protect people from gun violence. The BeSmart campaign was created to counsel families on firearm safety. BeSmart stands for: Secure guns in homes and vehicles, Model responsible behavior, Ask about unsecured guns in other homes, Recognize the risks of teen suicide, and Tell your peers to be SMART.

One of the main goals of Dr. Wright's visiting professorship was to identify and create community partnerships that will be dedicated to improving firearm safety in Hampton Roads. Following his time at CHKD, we felt that the formation of a strong partnership with Moms Demand Action and utilization of the BeSmart campaign would be the perfect first steps. We plan to advocate for firearm safety legislation at our capitol with MDA on February 4th and meet with student leaders at surrounding schools. We also have a spring goal to develop a quality improvement project aimed at increasing the number of firearm safety counseling sessions among providers in our resident General Academic Pediatric Clinic. We hope to implement a standardized firearm screening and counseling practice to be provided to all parents at well child visits.

We look forward to utilizing what we learned from our time with Dr. Wright and fostering our community partnerships as we embark on our advocacy goals. Preventing firearm violence is more critical now than ever, and **we owe it to our youth and our community to be leaders in working toward a solution.**



Children with untreated mental health needs are at increased risk for suicide, violence, and substance use. Nationally, one in five children has a diagnosable mental disorder and 1 in 10 suffers from a serious mental health problem that impairs how they function at home, school or in the community. Mental health problems that affect individuals throughout the lifespan typically have their onset in childhood: approximately 50% of psychiatric illnesses begin by age 15 and 75% begin by age 24 (Center for Behavioral Health Statistics and Quality (2016)). If children are not screened and treated, these childhood conditions may persist and lead to a cycle of school failure, poor employment opportunities, and poverty. In a five-year period, rates of severe youth depression have increased (Stagman, S. et al (2010). National Center for Children in Poverty).

In the State of Mental Health in America 2018 report, Virginia ranks 47th lowest in the country for mental health care for children under 18 years of age with 12.5% of children having had at least one major depressive episode (MDE) and 9.9% having had a severe major depressive episode. Virginia has the 7th highest rate of youth with alcohol dependence, 10th highest for marijuana use, 8th highest for cocaine use. Of Virginia's children with a MDE, 55,000 children (70.8%) did not receive mental health service. Of those who received treatment, only 15.5% received some consistent treatment. Virginia has a shortage of all mental health providers, ranking 42nd lowest in the country for the number of psychiatrists, psychologists, licensed social workers, counselors, therapists and advanced practice nurses specializing in mental health care per population (Nguyen, T. et al. (2018)).

Even more significant is the shortage of child psychiatrists in Virginia with only 0.13 child psychiatrist per 100,000 children in Virginia; 65% of the counties/cities in Virginia do not have a child psychiatrist at all and the remaining areas who have a child psychiatrist are in a severe or high shortage situation. The underserved regions of Virginia represent 98.8% of the state. Only two counties have sufficient numbers of child and adolescent psychiatrists, which represents only 23,086 of the 1.86 million children in Virginia (American Academy of Child and Adolescent Psychiatry).

Children and families that seek care from PCPs typically do not frame the visit as “mental health”–related (AAP Committee on Psychosocial Aspects of Child and Family Health and Task Force on Mental Health (2009)). Therefore, the role of community-based PCPs is unique and integral in a larger framework of providing a comprehensive mental health access program across a state. PCPs are in position to identify and understand the mental and behavioral health problems of their patients, even early in childhood. PCPs have over a dozen well attended preventative care visits with infants and young children and their families and these visits have “built in” primary care opportunities to ask sensitive questions about psychosocial adversity, cultural contributors, parenting stressors, as well as offer anticipatory guidance, brief behavioral interventions, home visits, and early intervention (Hagan JF, et al (2017) Bright Futures). Yet, in a recent national survey of more than 500 pediatricians, over 65% reported they lacked knowledge and skills in recognition and response to mental and behavioral health concerns. They lacked confidence in knowing when to refer or where to find resources for children and youth, let alone their parents (Cunningham PJ. (2009)). When mental health providers are available locally, wait lists are often long and referrals are not often completed (Kolko DJ (2009)).

The Virginia Chapter of the AAP is working to solve our mental health access crisis with a new program called the Virginia Mental Health Access Program (VMAP). When it is fully operational, the plan is to have four services operating in all regions of the state.

1. Primary Care Provider (PCP) education and training on screening, diagnosis, treatment and management of mental health disorders
2. PCP telephonic access to a Child Psychiatrist and Psychologist/Social Worker for consults (regional - north (Children's National/Inova), central (VCU), eastern (CHKD), western (UVA), southwestern (Carillion))
3. Telepsychiatry/telepsychology Services
4. Care Coordinators for each region to help find resources for families

We are working on advocacy for funding this program along with several other important child health initiatives.

I look forward to your help with advocacy and support so that we can improve our mental health services to children and adolescents in our state.

Sandy L. Chung, MD, FAAP, FACHE

President Virginia Chapter, American Academy of Pediatrics



VIRGINIA • PEDIATRICS NEWSLETTER ³

American Academy of Pediatrics – Virginia Chapter

Accreditation

This activity has been planned and implemented in accordance with the accreditation requirements and policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint providership of Eastern Virginia Medical School and the American Academy of Pediatrics-Virginia Chapter. Eastern Virginia Medical School is accredited by the ACCME to provide continuing medical education for physicians.

Credit Designation

Eastern Virginia Medical School designates this enduring material for a maximum of *1 AMA PRA Category 1 Credits™*. Physicians should only claim credit commensurate with the extent of their participation in the activity.

Content Director

C. W. Gowen, Jr., MD

Professor of Pediatrics, Eastern Virginia Medical School
EVMS Foundation Director
Chairman, Department of Pediatrics, EVMS
Senior Vice-President for Academic Affairs, CHKD

CME Committee

Kamil Cak, DMin, C.W. Gowen, Jr., MD,
Eric Y. Gyuricsko, MD, John Harrington, MD, Janice Karr,
Jamil Khan, MD, Windy Mason-Leslie, MD, Amy Perkins, Amy Sampson, Natasha Sriraman, MD

How to Obtain Credit:

Review the articles on pages 4-8. 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/VAAAPWinter2019> and complete online. Please allow 1-2 weeks to receive your certificate.

Disclosure of Significant Relationships with Relevant Commercial Companies/Organizations

Eastern Virginia Medical School endorses the ACCME Standard for Commercial Support that providers of continuing medical education activities, planning committee and speakers of these activities disclosure relevant relationships with commercial companies whose products or services are discussed in educational presentations.

For providers, significant relationships include large research grants, institutional agreements for joint initiatives, substantial gifts or other relationships that benefit the institution. For speakers, significant relationships include receiving from a commercial company research grants, consultancies honoraria and travel, other benefits, or having a self-managed equity interest in a company.

The following faculty have disclosed that they do not have an affiliation with any organization that may or may not have an interest in the subject matter of this CME activity and/or will not discuss off-label uses of any FDA approved pharmaceutical products or medical devices.



The CME committee members and content director have disclosed that neither they nor their spouses or partners have an affiliation with any corporate organization that may or may not have an interest in the subject matters of this CME activity.

Advantage of Screening Infant Hearing with aABR vs OAE

L. Ashleigh Greenwood, Au.D. | Clinical Director of Audiology for Pediatrix Audiology Services,
an affiliate of MEDNAX Services, Inc. | Inova Children's Hospital, Falls Church, VA
Chair of the Virginia Early Hearing Detection and Intervention (EHDI) Advisory Committee

Objective: To bring awareness to advantage of screening infant hearing with aABR vs OAE to avoid missing neural/brainstem hearing loss.

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



The goal of newborn hearing screening is early identification of hearing loss and subsequent timely enrollment in early intervention in order to prevent language and academic deficits. The latest research supports that those infants who meet the Early Hearing Detection and Intervention (EHDI) 1-3-6 criteria (screen hearing by 1 month of age, diagnose hearing loss by 3 months of age, and enrollment in early intervention by 6 months of age) have better language outcomes [1]. Therefore, newborn hearing screening with the most inclusive technology available is critical in early detection of hearing loss.

There are two methodologies currently in use for newborn hearing screening: Oto-acoustic Emissions (OAE) and automated Auditory Brainstem Response (aABR). An OAE screening assesses the outer and middle ear and can detect inner ear hearing loss related to cochlear outer hair cell dysfunction. Similar to OAE screening, aABR screenings assess outer ear, middle ear, and inner ear function, as well as the auditory nerve and auditory brainstem integrity. Automated ABR screenings will detect auditory nerve and/or auditory brainstem anomalies that OAE screenings would otherwise miss, as OAEs do not evaluate the auditory pathway beyond the inner ear (cochlear outer hair cell function).

Due to the ability for aABR screenings to detect auditory nerve and auditory brainstem anomalies, the most recent position statement from the Joint Committee on Infant Hearing (JCIH) in 2007 recommends using aABR as the primary screening method in the Neonatal Intensive Care Unit (NICU), as infants admitted to the NICU are at a higher risk for neural or brainstem anomalies [2].

One such anomaly is Auditory Neuropathy Spectrum Disorder (ANSD) which is a type of hearing loss that may affect: 1) cochlear inner hair cells; 2) the junction between the inner hair cells and the auditory nerve; or 3) the auditory nerve itself. An infant with ANSD may have largely intact cochlear outer hair cell function and would "pass" a newborn hearing screening when completed utilizing OAE methodology, thereby causing a delay in identification of the congenital hearing loss. Infants diagnosed with ANSD have a wide variety of functional hearing abilities; therefore, hearing loss resulting from ANSD that is missed on a newborn hearing screening may otherwise remain unidentified until early childhood when speech, language and learning deficits arise. An infant with ANSD is unlikely to pass an aABR screening, as this methodology relies on an intact auditory pathway from outer ear to brainstem; ANSD manifests itself in the part of the auditory pathway that is evaluated by aABR screening. Historically, approximately 1 in 10 cases of permanent hearing loss are believed to be ANSD [3]. However, recent research from well-born nurseries (WBN) suggests a significance prevalence of ANSD in this population. Boudewyns et al (2016) suggests an ANSD prevalence of 0.09 per 1000 babies in the WBN [4]. Among infants who referred on the newborn hearing screening in the WBN, Gerstenberger et al. (2018) indicated that a substantial amount of infants who referred on the newborn hearing screening via aABR were identified with ANSD in the WBN (81/127) versus in the NICU (46/127) [5].

At Inova Children's Hospital, Pediatrix utilizes aABR to screen both WBN and NICU infants, as auditory nerve and/or auditory brainstem anomalies are not limited only to infants who have been admitted to the NICU. Additionally, at Inova Children's Hospital, if a NICU baby refers on two hearing screenings prior to discharge from the hospital, a diagnostic ABR evaluation will be performed by a Pediatrix audiologist at the bedside, prior to discharge. The advantage of early detection of hearing loss is to provide parents with time sensitive information regarding their infant's hearing in order to enable parents to make the best decision for their family regarding the course of care. Subsequent enrollment into early intervention is desired in order to give the infant access to the best language outcomes. Delayed diagnosis, treatment, and intervention of hearing loss can have devastating effects on developmental speech and language milestones, literacy, and academic success.

If you have an infant or child in need of a newborn hearing screening, or a diagnostic audiologic evaluation, you may refer families to www.EHDipals.org to find a location near the family which provides these professional services.

References

[1] Yoshinaga-Itano C, Sedey A, Wiggin M, Mason C. Language outcomes improved through early hearing detection and earlier cochlear implantation. *Oto & Neuro*. 2018; 39:1256-1263

[2] Joint Committee on Infant Hearing. (2007). Year 2007 position statement: Principles and guidelines for early hearing detection and intervention. Available from www.asha.org/policy.

[3] Sininger YS. Identification of auditory neuropathy in infants and children. *Sem hear*. 2002; 23(3):193-200

[4] Boudewyns A, Declau F, van den Ende J, Hofkens A, Dirckx S, Van de Heyning, P. Auditory neuropathy spectrum disorder (ANSD) in referrals from neonatal hearing screening at a well-baby clinic. *Eur J Pediatr*. 2016; 175: 993-1000

[5] Gerstenberger J, Perez Abalo M, Brennhofner K, Porter K. (2018, March). Auditory Neuropathy Spectrum Disorder (ANSD) identified in Well-Baby Nursery (WBN) infants who referred newborn hearing screening. Poster presented at the 17th Annual Early Hearing Detection & Intervention Meeting, Denver, CO.



Tongue ties and upper lip ties: Why do they matter?

Kaelan Black, MD,

Ryan Murray, MD

Pediatric ENTs | Otolaryngology Associates, PC

Objective: Discuss the current research on tongue tie and lip tie.

ACGME Competencies: Patient Care, Practice-based Learning Improvement

In recent years the diagnosis of tongue tie, has reached “epidemic” levels with diagnosis of ankyloglossia (i.e. tongue tie) increasing over 800% from 1997 to 2012^[1]. This recent focus on the diagnosis of tongue tie is likely multifactorial and related both to an increased awareness of the benefits of breastfeeding along with more widespread breastfeeding resources that focus on assisting new mothers troubleshoot challenges in nursing [2]. The goal of this increased focus on ankyloglossia is part of an effort to improve rates of breastfeeding which has demonstrated benefits both important and diverse. Breastfeeding is associated with decreased rates of some infectious diseases (e.g. otitis media), lowering the risk of SIDS (sudden infant death syndrome), a decreased risk of childhood obesity, a decreased risk of breast cancer in nursing mothers, as well as lowering the economic burden of caring for a new child. Improving the latch of a child with ankyloglossia can significantly improve success in breastfeeding and directly decreases the rate of attrition in nursing mothers.

Ankyloglossia, the tethering of the tongue leading to limitations in tongue mobility, can occur at multiple anatomic locations^[3]. Mothers of infants with tongue tie will often complain of sore flat nipples, ulcers, difficulty latching or staying latched, clicking sounds, and increased air intake with feeding^[4]. Pediatricians, lactation consultants, and feeding specialists have been instrumental with diagnosing this problem and referring for effective intervention. For many decades, the “clipping” of a child’s tongue tie has been a widely accepted means to improve the latch of the infant struggling with nursing. This has been most frequently performed in children with “anterior” tongue ties, or a frenulum that attaches to the tip or mid tongue^[3]. With the release of these ties, mothers frequently report an immediate relief in pain with breastfeeding^[5]. “Posterior” tongue ties have traditionally been more difficult to diagnose and their association with breastfeeding difficulties is more controversial. This type of tongue tie is characterized by a lingual frenulum that inserts in the posterior portion of the mobile

tongue or just at its base, and leads to a restriction of the upward movement of the tongue^[3]. While some studies demonstrate significant improvement in feeding following release of a posterior tongue tie others have been less conclusive^[4-6]. Unfortunately, for all types of tongue ties, the recent increased focus on diagnosis has not yet been associated with high level studies examining outcomes following procedures to address these anatomic restrictions^[5, 6].

In addition to being associated with latch problems during breastfeeding, ankyloglossia may lead to difficulties in speech^[7]. Anterior tongue ties in particular should also be considered in cases of specific speech difficulties. A tethered anterior portion of the tongue may lead to difficulties with articulation of certain words and the release of this restriction has been shown to improve these speech problems^[7]. Following a release for speech difficulties, it is critical that children continue to work closely with a speech therapist to achieve optimal speech outcomes.

Ankyloglossia may also play a role in craniofacial development. The craniofacial impact of a tongue tie is theorized to include high and narrow palate, anterior and posterior crossbites, disproportionate growth of the mandible, and abnormal growth of the maxilla^[8]. These features have also been associated with sleep apnea due to narrowing of the pharyngeal airway^[9]. But like the other impacts of ankyloglossia, future high-quality studies are needed to better investigate these correlations.

The upper lip frenulum is another entity that has recently garnered considerable attention. If the upper lip is severely restricted, it may decrease the ability of the infant to appropriately latch^[4]. It has been suggested that while the majority of children are born with some degree of upper lip tethering, not all pose functional concerns. A critical factor to observe is whether or not the upper lip curls under itself during the latch. If this is observed in the context of breast feeding difficulties, a tethered upper lip frenulum may be implicated^[4].

Frenulectomy, or the release of the tongue tie, is a relatively safe and quick procedure that can be performed in the office in most infants under six months of age. If the child is older, the procedure is performed under anesthesia. There are two methods that are commonly employed to perform in-office tongue tie release. The first uses scissors to incise the frenulum after which the infant is immediately able to feed. Scissors have minimal bleeding during the procedure and cautery is rarely necessary. Scissors can also be used for release of an upper lip frenulum, but cautery is more often employed. The use of intraoral lasers has also become popular, especially among dental professionals. There are currently no studies demonstrating one method holds an advantage over the other. Instead, the factors that lead to the best outcomes following the release of tongue ties or upper lip ties, is the experience of the person performing the procedure and an accurate understanding of the etiology of each child’s specific feeding difficulties.

References

1. Douglas, P., *Untangling the tongue-tie epidemic. The Medical Republic*, 2017: p. 1-12.
2. Segal, L.M., et al., *Prevalence, diagnosis, and treatment of ankyloglossia: methodologic review. Can Fam Physician*, 2007. 53(6): p. 1027-1033.
3. Kollow, L., *Ankyloglossia (tongue-tie): A diagnostic and treatment quandary. Quintessence Int*, 1999. 30: p. 259-262.
4. Ghaheri, B.A., et al., *Breastfeeding improvement following tongue-tie and lip-tie release: A prospective cohort study. The Laryngoscope*, 2016. 127(5): p. 1217-1223.
5. Webb, A.N., W. Hao, and P. Hong, *The effect of tongue-tie division on breastfeeding and speech articulation: A systematic review. International Journal of Pediatric Otorhinolaryngology*, 2013. 77(5): p. 635-646.
6. Patel, J., R.P. Anthonappa, and N.M. King, *All Tied Up! Influences of Oral Frenulae on Breastfeeding and their Recommended Management Strategies. Journal of Clinical Pediatric Dentistry*, 2018. 42(6): p. 407-413.
7. Messner, A.H. and M.L. Lalakea, *The Effect of Ankyloglossia on Speech in Children. Otolaryngology-Head and Neck Surgery*, 2002. 127(6): p. 539-545.
8. Huang, Y., Quo, S., Berkowski, J., Guilleminault, C., *Short Lingual Frenulum and Obstructive Sleep Apnea in Pre-Pubertal Children. International Journal of Pediatric Research*, 2015. 1(1).
9. Guilleminault, C., S. Huseni, and L. Lo, *A frequent phenotype for paediatric sleep apnoea: short lingual frenulum. ERJ Open Res.*, 2016. 2(3).

Antinuclear Antibodies: When To Test And When To Consider Referral To Rheumatology

Bitra Arabshahi, MD

Medical Director of Rheumatology

Pediatric Specialists of Virginia

Objective: To provide guidance to general pediatricians regarding ANA testing and interpretation.

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

While Antinuclear Antibodies (ANA) play an important role in many autoimmune disorders, their poor specificity and high prevalence pose a dilemma when it comes to decisions about ordering and interpreting this test. It is estimated that approximately 30% of healthy individuals have positive ANA titers in the 1:40 dilution, 13% in the 1:80 dilution, and 5% in the 1:160 dilution⁽¹⁾. In order to guide testing/referrals and alleviate patient anxiety stemming from positive ANA results, the following guidelines may be of value to pediatricians.

When Should I Order an ANA?

There are three situations in which is reasonable to order ANA testing. The 1st situation is in cases where you clinically suspect that your patient may have lupus or other connective tissue disease. In such cases, one would expect to have objective findings, such as a malar rash, serositis, myositis, arthritis (especially in high-risk demographics such as African Americans, Hispanics or Asians), or unexplained cytopenia or proteinuria in the setting of elevated inflammatory markers. The second situation is in patients with Raynaud's phenomenon. Raynauds in the setting of a negative ANA and normal nail fold capillaries is considered Primary Raynauds, and warrants reassurance and a pair of gloves. Raynauds in the setting of a positive ANA often warrants further investigation as it is often secondary to other autoimmune conditions such as lupus, scleroderma, Mixed Connective Tissue Disease, or antiphospholipid antibody syndrome⁽⁴⁾. The third situation in which ordering ANA testing is appropriate is in the patient with known or suspected Juvenile Idiopathic Arthritis (JIA). It is important to understand that a positive ANA result in JIA has no correlation with the presence of arthritis, but it is correlated with a higher risk of uveitis in JIA patients. ANA presence in JIA can thus help guide our recommendations regarding the frequency of ophthalmologic screening in this population⁽³⁾.

How Should I Order an ANA?

Immunofluorescent antinuclear antibody test using HEp-2 cells (ANA by IFA) is considered the gold standard for autoantibody detection, and specific autoantibodies are generally tested by ELISA as a next step. It is important to order quantitative ANA titers rather than qualitative screens, as screens can sometimes be reported as positive with very low titer/insignificant ANAs. In cases where a specific connective tissue disease such as lupus or scleroderma is suspected, ordering an "ANA titer by IFA with reflex to cascade" will prompt the lab to process dsDNA and extractable nuclear antigens (ENA), including anti Smith, SSA, SSB, RNP and Scl70. These results, along with presence or absence of clinical symptoms, can determine if the positive ANA is of clinical significance.

What Should I Make of ANA titers and patterns?

While there is some truth in saying that higher titers of ANA are more clinically significant, transient elevations in ANA titers in children with infectious diseases or self-limited autoimmune disorders such as Henoch Schoenlein Purpura, is common. Presence of ANA in the serum can also be associated with genetic factors (through histocompatibility locus DR3), environmental agents (drugs, particularly those associated with drug-induced lupus, such as minocycline), other autoimmune conditions such as hypothyroidism or vitiligo, and neoplasms. In absence of any objective clinical and laboratory findings concerning for rheumatic disease, checking and following ANA titers is not necessary, regardless of the titer. One exception may be in cases where ANA titers are greater than 1:1280. Persistent elevated ANA tiers in this range can be associated with future risk of scleroderma and warrant followup.

The ANA profile, or ENA, often provides more specific guidance regarding the presence of an autoimmune condition, although false positive ENAs can also occur. The presence of dsDNA and/or anti Smith antibodies is associated with lupus; SSA/SSB antibodies are often seen in the setting of Sjogren's Syndrome; Scl-70 positivity is seen with scleroderma, Jo-1 positivity is sometimes seen with Dermatomyositis, and anti-histone antibodies in absence of dsDNA/anti Smith are associated with drug-induced lupus⁽²⁾.

With regard to the ANA pattern, findings are generally non-specific. Although homogenous or speckled patterns are often reported by the lab as being associated with lupus, these patterns are seen equally as often in healthy individuals with clinically insignificant ANAs. ANA patterns may serve as a guide to rheumatologists, but are not diagnostic of any particular disease. What makes an ANA significant, is associated objective clinical findings. Future Directions: The presence of a novel antibody, anti DFS70, seen in positive ANAs with a dense fine speckled pattern, may serve as a good marker for the absence of rheumatic disease. Up to 22% of individuals with DFS70 positive ANAs were found to be healthy, while this antibody was only seen in 1% of ANA positive individuals with rheumatic disease⁽⁵⁾. Further confirmatory studies and future commercial availability of this test may assist in referral of patients found to have positive ANAs.

References:

- (1)Tan EM, Feltkamp TE, Smolen JS, Butcher B, Dawkins R, Fritzler MJ, et al. Range of antinuclear antibodies in "healthy" individuals. *Arthritis Rheum.* 1997;40:1601-11.
- (2)Sato M, Chan EKL, Sobel ES, Kimpel DL, Yamasaki Y, Narain S, et al. Clinical implication of autoantibodies in patients with systemic rheumatic diseases. *Exp Rev Clin Immunol.* 2007;3:721-38.
- (3)American Academy of Pediatrics Section on Rheumatology and Section on Ophthalmology: guidelines for ophthalmologic examinations in children with juvenile rheumatoid arthritis. *Pediatrics* 1993;92:295-6.
- (4)Kallenberg, CG, Pastoor GW, Wouda AA et al. Antinuclear antibodies in patients with Raynaud's phenomenon: clinical significance of anticentromere antibodies. *Ann Rheum Dis.* 1982; 41:382-387
- (5)Conrad K, Rober N, Andrade LE, Mahler M. The Clinical Relevance of Anti-DFS70 Autoantibodies. *Clin Rev Allergy Immunol.* 2017;52:202-216

Craniosynostosis

Stephen B. Baker, MD, DDS

Medical Director Craniofacial Anomalies Program | Inova Children's Hospital

Objective: *Bullet overview of craniosynostosis*

ACGME Competencies: *Patient Care, Medical Knowledge*

Craniosynostosis is defined as the premature closure of a cranial suture which causes abnormal calvarial growth. Preoperative assessment for craniosynostosis includes a detailed medical history, physical examination, and radiographic imaging.

Surgical intervention is indicated in craniosynostosis both for the correction of calvarial contour deformities and the prevention of psychosocial dysfunction, intracranial hypertension, and/or mental retardation. Studies have shown that the presence of intracranial hypertension is dependent on the number of affected sutures, ranging from approximately 14% for single-suture synostosis to approximately 47% in multisuture synostosis.^{1,2} Sutural release in simple craniosynostosis has been advised due to the concerns regarding increased intracranial pressure as well as the mild but significant developmental delay in the aging child. Patients with complex synostoses present with increased severity of physical and neurological symptoms; therefore, surgical intervention is even more imperative.

PATIENT PREPARATION

An open craniofacial approach remains the mainstay of therapy, relying on wide scalp dissection, extensive calvarial osteotomies, and skull reconfiguration that is individually tailored to each cranial vault deformity.³ To address concerns regarding incision length, operative blood loss, and length of stay for open craniofacial procedures, minimally invasive techniques have been proposed. These techniques include endoscopic sutural release, spring-assisted cranioplasty, and distraction osteogenesis.^{4,5,6}

The majority of craniofacial surgeons operate between 3 and 12 months of age. Minimally invasive techniques, which rely on dynamic cranial vault alteration during rapid calvarial growth, are generally performed at an earlier age than open surgical correction.

COMPLICATIONS

Perioperative morbidity may include wound infection bleeding, dural laceration, superficial brain injury, cerebrospinal fluid leak, encephalocele formation, subgaleal hematoma, and ocular injury. Postoperative mortality rates are low and continue to decline with technological advancements and experience.

REFERENCES

1. Renier D, Sainte-Rose C, Marchac D, Hirsch JF. Intracranial pressure in craniosynostosis. *J Neurosurg.* 1982;57(3):370-377.
2. Thompson DN, Harkness W, Jones B, et al. Subdural intracranial pressure monitoring in craniosynostosis: its role in surgical management. *Childs Nerv Syst.* 1995;11(5):269-275.
3. Whitaker LA, Munro IR, Salyer KE, et al. Combined report of problems and complications in 793 craniofacial operations. *Plast Reconstr Surg.* 1979;64(2):198-203.
4. Jimenez DF, Barone CM, Cartwright CC, Baker L. Early management of craniosynostosis using endoscopic-assisted strip craniectomies and cranial orthotic molding therapy. *Pediatrics.* 2002;110(1 Pt 1):97-104.
5. Lauritzen CG, Davis C, Ivarsson A, et al. The evolving role of springs in craniofacial surgery: the first 100 clinical cases. *Plast Reconstr Surg.* 2008;121(2):545-554.
6. Akai T, Iizuka H, Kawakami S. Treatment of craniosynostosis by distraction osteogenesis. *Pediatr Neurosurg.* 2006;42(5):288-292.



Does My Child Have an Arrhythmia?

Mitchell Cohen MD, FACC, FHRS

Co-Director of the Pediatric Heart Program | Director of Pediatric Electrophysiology

Inova Fairfax Children's Hospital

Professor of Pediatrics

VCU School of Medicine

Objective: Understand clinical symptoms and when to consider catheter ablation.

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



As a pediatric electrophysiologist I am often faced with the challenge of trying to determine if a child is having an arrhythmia. The evaluation of the patient suspected of having an arrhythmia necessitates understanding whether the palpitations start and stop abruptly or have a more gradual onset and termination. The abrupt onset and termination are consistent with a reentrant circuit and likely supports either an accessory pathway (i.e. WPW), AV nodal reentrant tachycardia, or atrial flutter (the latter is more common in patients with congenital heart disease). In these patients' efforts should be made to use some sort of remote rhythm monitor to capture an event. Reentrant SVT can start during exercise, post-exercise, or at rest. The ventricular rate of a reentrant circuit is generally 190-240 bpm and shows a narrow QRS complex often with retrograde P waves. Tachycardias that have a more gradual onset and termination ("warm up and cool down") are often sinus tachycardia and related to stress, anxiety, or other causes for adrenergic surges. However, other automatic tachycardias include ectopic atrial tachycardias (EAT) that have an atrial focus different than the usual sinoatrial node. These patients are a little more challenging to diagnose and often the rates of the tachycardia are

120-150 bpm. It is important that patients with EAT are diagnosed early as the chronic elevated heart rates may result in a cardiomyopathy. Patients with frequent PVCs (>25-30% per day) may also be at risk for developing a cardiomyopathy. If these patients develop echocardiographic findings of decreased left ventricular function or symptoms associated with the PVCs, anti-arrhythmic therapy or catheter ablation should be considered.

Beyond kindergarten the likelihood of outgrowing the SVT becomes less. While anti-arrhythmic medications are often reasonable to consider in the young child, the reality is that such an approach is only palliative. The side effect profile for anti-arrhythmic drugs remains high, noncompliance is an ongoing issue especially amongst adolescents, and medications do not absolve the patient of SVT episodes. The only permanent treatment for patients with SVT is catheter ablation. The indications for catheter ablation include patients with SVT refractory to anti-arrhythmic medications, patients with pre-syncope or syncope associated with palpitations, a desire to be off medications, or concern for a life-threatening arrhythmia and sudden cardiac death. The latter is especially true in patients with Wolff-Parkinson-White syndrome who may be at risk for rapidly conducting atrial fibrillation and carry a 1-2% risk of sudden death over one's lifetime.

Catheter ablation has evolved over the last two decades and is now the preferred treatment option for most patients beyond the age of 8. The ablation is performed either under conscious sedation or general anesthesia depending on the age and desires of the patient. Once the tachycardia is induced in the EP laboratory, the abnormal circuitry is mapped using a highly complex three-dimensional mapping system that recreates an anatomic shell of the endocardial surface of the myocardium and can identify the earliest abnormal electrical focus that is causing the SVT. Using the latest automated three-dimensional technology, I have the ability in a matter of 30-45 minutes to obtain 500-1,000 voltage activation points to accurately localize the source of the SVT within millimeters. Once the tachycardia has been accurately mapped I may use radiofrequency energy ("heat") to create a small scar over the focus or utilize cryoenergy ("freezing") to ablate the focus. The decision to use heat or freezing generally rests with an understanding of where the abnormal electrical circuit resides in proximity to the normal native AV conduction. If the location of the SVT circuit is too close, within 4-6 mm, of the native AV conduction tissue I will use cryoenergy as it is generally safer than RF energy with a significantly lower risk of damaging the normal AV node. The success rate of catheter ablation for SVT in children and adolescents is 94-95%. Complications of catheter ablation are rare (<1%) but do include injury to the native conduction tissue, vasculature, or coronary arteries. As always, a careful discussion between the electrophysiologist, patient, and parents must go through the risk-benefits of the procedure versus medication or observation. Children are usually restricted from sports participation for 1-week post procedure and then may resume a full complement of activities without restrictions and no need for anti-arrhythmic medication. We have looked at the quality of life for children and adolescents following a successful ablation for SVT and have observed a tremendous improvement in their overall well-being and perception of health and happiness. In 2018, I performed 120 ablations at Inova Fairfax Children's Hospital for children with SVT or VT with a success rate of 96.5%. The future of pediatric arrhythmias in children remains excellent and most patients can be cured without the need for life-long medical therapy and require no activity restrictions after catheter ablation.

Dr. Mitchell Cohen sees patients at both the Pediatric Cardiology Associates and Child Cardiology Associates offices as part of the Inova Electrophysiology Program. (Office Number 703-942-8300)

VA-AAP Newsletter Registration and Evaluation Form (Winter 2018)

You have the opportunity to claim up to 1 AMA PRA Category 1 Credit(s)[™].

To claim CME credit, please complete the survey below.

Name: _____ Degree: _____

Last four digits of SSN: _____ Email address: _____

Mailing address: _____

For this activity, how many hours of CME are you claiming? _____ (Max. 1.5 hours)

As a result of reading the articles, will you make any changes in your practice? Yes No

Please list at least 3 strategies that you plan to implement as a result of reading the articles? *(answer required for credit)*

1. _____

2. _____

3. _____

If you will not make any practice changes, did this activity reinforce your current practice of pediatrics? Yes No

Please explain:

How could this activity be improved?

Future Topic Requests (optional):

Overall, how would you rate this activity?

Excellent		Average		Poor
5	4	3	2	1

This CME activity will expire on November 2019.

Please send completed form to

EVMS-CME

358 Mowbray Arch #207

Norfolk, VA 2350

2019 Pediatric General Assembly Day
January 17, 2019
Virginia Chapter Members Advocating for Kids!



American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN™
Virginia Chapter



2019 Legislative Priorities

Access for Children and Adolescents to Mental Health Services

Support Governor's Budget Item 311 #F, \$1.23 million

Virginia Mental Health Access Program (VMAP)

- There is a severe shortage of child psychiatrists in Virginia; only two counties have a sufficient number
- Statewide mental health access program for children designed to give children's healthcare providers access to child psychiatrists, psychologists, social workers & care coordination
- Provides more children with mental health services by expanding the existing workforce

Ensuring Children's Access to Health Care Services

Support SB1344 (Favola)

Ensuring all Health Plans include Essential Health Benefits

- Two new types of insurance plans are being advanced by the federal government—short-term, limited duration (STLD) plans and Association Health Plans (AHPs) that are not required to offer EHB. While such plans might be cheaper upfront, they can also provide very little coverage—which families might not realize until they need it.
- EHB are critically important for children, and include maternity and newborn care; mental health and substance use disorder services; pediatric services, including oral and vision care; prescription drug coverage; and preventive care

Reducing Children's Risk of Firearm Injury

Support HB2206 (Filler Corn)

Sales Tax Exemption; Gun Safes

- Creates retail sales tax exemption for purchase of gun safe with a selling price of \$1,000 or less
- Unintentional injuries are the leading cause of death in children older than 1 year. Minimizing the risk of injury, including locking up firearms, plays a key role in injury prevention.
- About 1/3rd of U.S. children live in homes with firearms and 43% of these contain at least 1 unlocked firearm.

Support HB1763 (Sullivan)/ SB1458 (Barker)

Firearms; Removal from Persons Posing Substantial Risk

- This will close a gap in VA law by giving law enforcement the tools they need to temporarily remove firearms from individuals who pose a substantial risk of personal injury to self or others but who do not meet the current requirements for removing a firearm as outlined in § 18.2-308.1:3
- This will give parents and citizens a tool when they believe someone poses a threat, especially to children, in their communities.

Finding Solutions for Food Insecurity and Obesity

Support SB999 (Stanley)/ SB1189 (Wagner)/ HB1858 (McQuinn)

Virginia Grocery Investment Fund

- VGIF would create a public-private partnership leveraging state dollars with private money that will provide one-time, low-interest loans to encourage grocers to open or renovate stores in underserved communities.
- An investment of \$5 million from the state could leverage \$15 million in private capital, creating a \$20 million dollar Grocery Investment Fund which could support over 10 healthy food retail projects in Virginia.



VIRGINIA • PEDIATRICS

2018 Child Advocate Award Presentation



Each year, the Virginia Chapter of the AAP recognizes an individual(s) in our Commonwealth who stand up for the rights, values, and recognition of Virginia's children. Sandy Chung, MD, VA-AAP President presented Dr. Bela Sood with the 2018 Virginia Chapter, American Academy of Pediatrics Child Advocate Award in recognition for all her support of many of the Virginia Chapter's proactive initiatives as well as her tireless advocacy work throughout the years. Our Virginia Chapter members wanted me to personally thank Dr. Sood, for your significant contributions to healthcare for Virginia's children.

Dates to Remember!

Clinical Challenges in Pediatric Primary Care 2016

March 23, 2019

Lewis Ginter Botanical Garden

1800 Lakeside Avenue

Richmond

Contact: Sherry Black (804) 828-4790

McLemore Birdsong Conference

Omni Charlottesville

April 12 –14, 2019

For more information and registration go to www.cmevillage.com

2019 Peds at the Beach Conference

July 27 – 29, 2019

Hilton Oceanfront

Virginia Beach

Questions? Contact VCU Health Continuing Medical Education
cmeinfo@vcuhealth.org | 804.828.3640