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President's Letter

The Year in Review from a Child Health Policy Perspective: Making Lemons from Lemonade



Helen DuPlessis, MD, MPH, FAAP
President, AAP California, Chapter 2

I generally enjoy one or two well-conceived, journalistically accurate “year-in-review” offerings at the end of each calendar year. I suspect that is because those retrospectives tend to be uplifting and affirmative even when the year has had more than a smattering of tragedy. But for some unknown reason, I managed to miss (or unconsciously avoid) all of them at the end of 2012. So, in contemplating the topic for the Chapter President’s newsletter, I thought I might construct my own year in review – through the lens of child health providers and advocates, with an eye toward the positive. While I certainly will not presume this offering to be as moving as some, I hope it is at least an accurate accounting, and perhaps even a cause for reflection.

In the state of California

Even as we grieved from children killed and injured in shootings in Aurora, Colorado; Portland, Oregon; Oak Creek, Wisconsin; and in Newtown, Connecticut; fewer children died from firearms in California than in recent years. While a child or teen dies or is injured from a gun every 30 minutes in the United States, our country is engaging in serious public debate about reducing gun violence for the first time in centuries. President Obama released a set of gun violence prevention recommendations on January 16th. Let’s hope the 2013 retrospective will celebrate the signing of legislation incorporating all or most of those recommendations <http://www.whitehouse.gov/issues/preventing-gun-violence>

CALENDAR OF EVENTS

AAP-CA2

January 26 - Saturday

**2013 Pediatric Surgery
Conference**

9am - 4:15pm,

Children's Hospital Los Angeles

Information & RSVP

www.CHLA.org/

[PedSurgeryConference](#) or

contact Caitlin Beck at

cbeck@chla.usc.edu

January 31 - February 1

3rd Annual California

Breastfeeding Summit

More information is on our

website, including registration

link. <http://aapca2.org/events>

February 4 - Monday

Speak Up for Children at the

Federal Level - Advocacy

Training

8:00am - 5:00pm,

Washington DC

Contact Jamie Poslosky at

jposlosky@aap.org

February 5 - Tuesday

Ventura County Children's

Oral Health Summit

"Great Health Starts Here"

8:00am - 2:00pm,

Camarillo, California

Contact Susan Englund at

[805-485-6288](tel:805-485-6288) x 226

February 6 - Wednesday

Town Hall:

"The Heart and Minds

of Children" focusing on

concussion management

6:30pm - 9:00pm,

Bistro Garden

12950 Ventura Blvd

Studio City

RSVP to Ken Saul at

docsaul@aol.com

March 7 - Thursday

Town Hall: "Acne

Management"

Santa Monica

Contact Corinn Cross at

coricross@hotmail.com

An extension of the California Child Passenger Safety Seat law went into effect requiring children under 8 years of age or under 4'9" tall to be restrained in the back seat using a car seat or booster, as appropriate.



Moreover, hospitals, clinics and birthing centers must inspect the car seat prior to discharging mothers and newborns, and provide information about obtaining low cost car seats.



AB 2109, the Immunization Vaccine Education act was signed into law, requiring parents/guardians seeking Personal Belief Exemptions (PBE) from vaccinating school-aged children to demonstrate they have discussed the pros and cons of immunization with a physician or other eligible provider by presenting a signed letter documenting that education was given. The bill was authored by our own Richard Pan, MD, FAAP, and signed into law despite well-organized opposition from anti-vaccine protestors. The steady rise in PBEs in recent decades (concomitant with the periodic measles, and more frequent pertussis epidemics) largely results not from parents/guardians who object to vaccination, but more often from parents/guardians who, for whatever reason, are rushing to tie up all of the loose ends required to enroll children into public schools, and sign a PBE to expedite the school enrollment process (sometimes wrongly encouraged to do so by well-meaning helpers). Let's hope this legislation, along with the expanded access to health insurance for California's children and families, will reverse or at least halt that trend. Many colleagues are dreading the onslaught of parents seeking PBE documentation, but I'm hopeful that few will be turned away, and that hospitalizations and deaths from vaccine preventable diseases will ebb once again.

A very weathered, and shrewd Governor Brown succeeded in persuading voters to approve Proposition 30, which increased the sales tax by ¼ of a cent and gave him the authority to raise property taxes on the wealthy (i.e., earning over \$250,000/year). The revenue generated is initially earmarked for K-12 schools and universities, offering some relief from the inexorable cuts to state spending on public schools over the past 3 years. Given the proven link between education, health and longevity, support for education is a necessary pathway to health.

After a reasonable effort at seeking advice from child health advocates and providers, the state began the phased transition of over 860,000 children previously covered by the Healthy Families into the Medi-Cal program. While it is too soon to tell how smoothly that transition is progressing, and understandable fears abound regarding potential disruptions in care, and administrative complexity, the move promises children the security of a more comprehensive benefit package, guaranteed coverage for eligible children (Medicaid is an entitlement while Healthy Families was not), and with the pending Medicare parity shifts for most pediatrics providers, slightly better reimbursement rates in the long run.

Clinical and Technological Advances

I'm not certain I have a bead on all of the clinical advances in pediatrics in 2012. While clearly important advances in HIV, cancer and respiratory therapy have occurred, these advances seem remotely hospital or research-based for now, and will require a little time for their importance to translate to our practices and communities. Speaking of hospitals, in 2012 the Centers for Medicare and Medicaid Services funded 26 Hospital Engagement Networks (HEN) to take on the task of reducing "hospital acquired conditions," and hospital readmissions in Medicare patients. Only one of the hospitals networks, Ohio Children's Hospital Solutions for Patients Safety, was a children's hospital network.

The older I get, the more I am concerned about health care for the elderly, but at the same time, I am equally convinced that our continued focus on improving care for the elderly, and those with chronic conditions – while important and necessary – misses a critical opportunity to focus on optimizing health and developing healthy behaviors at the earliest possible stages in the life course (pregnancy and early childhood). Focusing more on prevention in the very broadest sense, and primary care has been proven time and time again to defer or prevent many of the chronic conditions plaguing our elderly. I understand that this isn't sexy and won't realize the short term cost savings demanded by the ACA, but if we can't commit to an upstream strategy that begins with children, we have no hope of shifting the health care cost curve that is driven by chronic disease near the end of the lifespan.

The Affordable Care Act

Nearly 3 years after the Patient Protection and Affordable Care Act (ACA) was signed into law, more than half of the provisions have been put into effect including those you know well: prohibition of coverage denial for pre-existing conditions; coverage of dependents through 26 years of age; tax credits to small business that provide health insurance to their employees; Medicaid drug rebates; establishment of the Centers for Medicare and Medicaid Innovation; establishment of a prevention trust fund; elimination of consumer copayments on preventive services (in new plans).

In June, the ACA withstood a Supreme Court challenge, upholding nearly all of the controversial provisions (including the individual mandate, albeit with a justification of Congressional taxing authority, rather than its authority to regulate interstate commerce), excepting the withholding of Medicaid funds from states that refuse to implement the Medicaid expansions in the ACA.

January 24 - Thursday
ClzQIDS Vaccine Study
Recruitment - Webinar
12:00 pm Eastern
<https://cnmc.webex.com/cnmc/j.php?ED=204922852&UID=498639607&RT=MiMxMQ%3D%3D>

or to apply for project,
fill out application at
<https://cri-datacap.org/surveys/?s=gd3HoE>

Questions?
contact Liz Rice-Conboy, MS
at ericeconboy@aap.org

SAVE THE DATE...

Advances in Pediatrics AAP-CA2 CME

May 2nd - 5th 2013

Ceasars Palace Hotel &
Casino
Las Vegas, Nevada

SPEAKERS INCLUDE:

Francine Kaufman, MD
Endocrinology

Lawrence Eichenfield, MD
Dermatology

Martin Stein, MD
Behavioral and
Developmental Screening

Paul Krogstad, MD
Infectious Disease

Wilbert Mason, MD &
Helen DuPlessis, MD
Quality Improvement and
Maintenance of Certification

Registration will be opening
soon.

(\$50 off for previous attendees)

visit <http://www.aapca2.org>

for the latest news and
information.

As of the (extended) deadline in 2013, 20 states submitted applications to establish health insurance exchanges to facilitate the purchase of individual insurance plans by creating a purchasing exchange aimed at realizing lower cost options. While the other states are positioned to take advantage of an exchange(s) established by the federal government, there may still be challenges to surmount before that can happen. California, having some experience implementing insurance cooperatives in the past, has been a leader in making progress on its exchange, known as the California Health Benefit Exchange (HBEx). Still the process has been slow – here and elsewhere – and the learning collaborative that should have been established so that states could learn from each other rather than reinventing the wheel while implementing this complex provision, has not been operational. One would hope that involved individuals and organizations at the state and federal level would very quickly recognize the need for and encourage an unprecedented amount of cooperation and planning to optimize the success of these exchanges. There's still time to leverage this knowledge as the exchanges move from planning to implementation.

Although not many parents will need to seek coverage for their children through the exchange, a few will, and we are now in the throws of identifying an essential health benefit (EHB) package appropriate for children. That EHB must be designed to recognize the unique epidemiology of health and disease, growth and developmental needs of children, as well as the importance of prevention, screening and the development of healthy behaviors. The Kaiser Small Group product being considered by HBEx, while adequate for adults, falls short for children in a variety of ways. Conversations are ongoing with state officials about the California EHB, and with federal officials about the regulatory language guiding states' decision-making about EHBs.

Looking toward 2013

There is much that was accomplished in 2012, and much more to be done in the future. As we enter the 13th year of this 21st Century let us hope that leaders at every level are guided by clear principles that ensure infants, children and youth are supported to have their needs satisfied and realize their potential to interact with their environment and become contributing members of society. Moreover, I wish that their health care providers are similarly supported to care for them.

CAHAN ALERT

In the past several weeks, there has been a marked increase in Emergency Department visits in Los Angeles County (LAC) for respiratory illness.

Respiratory syncytial virus (RSV) prevalence has greatly increased over this time to the highest levels in 4 years. Influenza in LAC has also been gradually increasing and is expected to peak later in January or February. Please see the attached Flu Surveillance Update.

RSV is a respiratory virus that infects the lungs and breathing passages. RSV is a contagious viral disease that can lead to serious health problems-especially for young children and older adults. Prophylaxis with paliviumab may be considered for certain high risk infants and children. For more information on RSV and prevention go to: <http://www.cdc.gov/rsv/about/index.html>.

Because influenza has not peaked here in LAC, there is still time to vaccinate yourself, your staff, and your patients. LAC DPH, the CDC, and the Advisory Committee on Immunization Practices recommend that all persons 6 months of age and over, including healthy adults be vaccinated for influenza. It is especially important for the following groups to get a flu vaccine:

- Infants/ babies 6 months through 18 years of age
- Adults 50 years and older
- Pregnant women or women who have just had a baby
- Morbidly obese
- Living in a nursing or long-term care home
- Living with weakened immune system or chronic medical condition
- Living with or caring for someone who is more likely to have flu complications
- Healthcare providers

For information on where to receive a flu vaccine go to <http://publichealth.lacounty.gov/ip/flu/FluLocatorMain.htm>

RSV and influenza are not reportable diseases but all influenza deaths and suspected outbreaks of any kind are reportable. For more information please contact Acute Communicable Disease Control at (213) 240-7941

FOR YOUR INFORMATION...

Do you need more preservative-free flu vaccines for children under 3 yrs of age?

Sanofi, the manufacturer of preservative free flu vaccine for children under 3 has resumed production after it ran out at the beginning of January. They are taking orders for delivery at the end of January. In the mean time, our Chapter and District are working on getting a waiver to allow children under 3 to be vaccinated with regular flu vaccine now while the preservative free vaccine is unavailable.

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Updates in Autistic Spectrum Disorder: Diagnosis and Access to Services

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Autistic spectrum disorders (ASDs) are characterized by 1) deficits in communication, 2) impairments in social interactions, and 3) restricted and repetitive patterns of behavior (American Psychiatric Association, 2000). Under the current DSM-IV-R classification system, ASDs are considered in the category of pervasive developmental disorders (PDDs). This category includes Autistic Disorder, Rett's Disorder, Childhood Disintegrative Disorder, Asperger's Disorder, and Pervasive Developmental Disorder Not Otherwise Specified (Figure 1). There are proposed changes in the DSM as well as recent changes in the Regional Center and private insurance systems which are important for practicing pediatricians in California to know about. Other issues related to ASDs including specific therapies and medication management will be discussed in a future article.

On the issue of diagnosis, there are anticipated to be specific changes when DSM-5 is released in 2013. One of the more significant changes would be a modification in terminology that does away with the Asperger's Disorder classification. Studies that looked at different centers diagnosing autism found that the diagnosis given in specific cases was more dependent on the treatment center than the clinical presentation (Arch Gen Psychiatry. 2012; 306). However, the specific label which a child received could play a significant role in whether they received services (California Welfare and Institutions Code, Section 4512). Given these concerns about diagnostic specificity, it was felt that the categorization should be removed.

Another notable change with DSM-5 would involve combining the symptom categories of 1) deficits in communication with 2) impairments in social interactions. This change is being considered because of the significant overlap between the two categories. The third category of symptoms, restricted and repetitive patterns of behavior, would remain a distinct entity. A new diagnosis named social communication disorder would be used for a child with these deficits in communication and impairments in social interactions but without the restricted and repetitive patterns of behavior. This new diagnosis is proposed as a communication disorder as opposed to a pervasive developmental disorder (Figure 2). Another change regarding these behaviors will be the recognition of abnormal sensory responses as a type of restricted and repetitive patterns of behavior (DSM5.org).

In the new system, children with a PDD would also be classified using multidimensional categories including severity level (Figure 3), underlying genetic diagnosis (i.e. Fragile X), comorbid language, learning, psychiatric disorders (i.e. ADHD, depression), comorbid medical disorders (i.e. seizure disorder, IBS), and age/pattern of onset (i.e. with regression). It is important to be aware that these changes are not finalized yet. More information is available at www.dsm5.org.

Another area of importance for practitioners is behavior and other therapies for children with autism, and how to fund those therapies. Behavior therapy, specifically applied behavioral analysis (ABA), is an evidence-based, intensive intervention that is built on a conditioning and reinforcement model. Its methods are designed to “increase and maintain desirable adaptive behaviors, reduce interfering maladaptive behaviors or narrow the conditions under which they occur, teach new skills, and generalize behaviors to new environments or situations” (Developmental and Behavioral Pediatrics – Evidence and Practice. 2008; 549). ABA comes in different forms, including discrete trial training (DTT) and pivotal response training (PRT). DTT breaks tasks down into short simple trials and rewards desired behavior and task completion (Consult Pediatrics. 2010; 303). PRT involves reinforcement activities that build on learner initiative and interests, enhance motivation to respond to social cues, improve management of one’s own behavior and improve self-initiation of behavior in appropriate contexts (Journal of Positive Behavior Interventions. 2010; 23). For children who are higher functioning, PRT may be a more flexible and interactive model of behavior therapy.

In California, adaptive skill therapies like ABA have been historically funded through the Regional Center system. Children under 3 years old with developmental delay have been eligible for Regional Center services through the Early Start Program (California Government Code, Section 95014). Children with a diagnosis of autistic disorder as well as significant functional limitations have been eligible for Regional Center funding beyond 3 years old through the Lanterman Developmental Disabilities Act (California Welfare and Institutions Code, Section 4512). Since 2008, changes to eligibility for Early Start have influenced access to these services. Historically a 33% delay in any one category of developmental skill was necessary for Early Start eligibility. As of 2008, children 24-35 months old must evidence a 50% delay in one category or a 33% delay in two different categories. Regional Centers also now require clients to exhaust private insurance coverage for therapies prior to receiving Regional Center funding.

Another new provision affecting access to behavior therapy services for children in California with ASD is the new California Senate Bill 946. This new law, which took effect in July 2012 and sunsets July 2014, requires private health care service plans and health insurers to provide coverage for behavioral health treatment for pervasive developmental disorder or autism. It is notable that the bill’s coverage requirement includes a broader range of patients than have been historically covered by Regional Center services beyond 3 years old. By including children with PDD in general, children with less severe ASDs could be eligible for behavior therapy funding to support their development and functioning. This places a greater emphasis on proper diagnoses for children with autistic spectrum disorder by providers, including psychologists and developmental-behavioral pediatricians, to ensure insurance coverage.

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It remains to be seen how these changes will affect the prevalence of ASDs and access to services for these children. However, it will be important for pediatricians to stay informed to be able to help their patients with developmental issues receive proper services.

Figure 1 – DSM-IV-R classification of PDDs

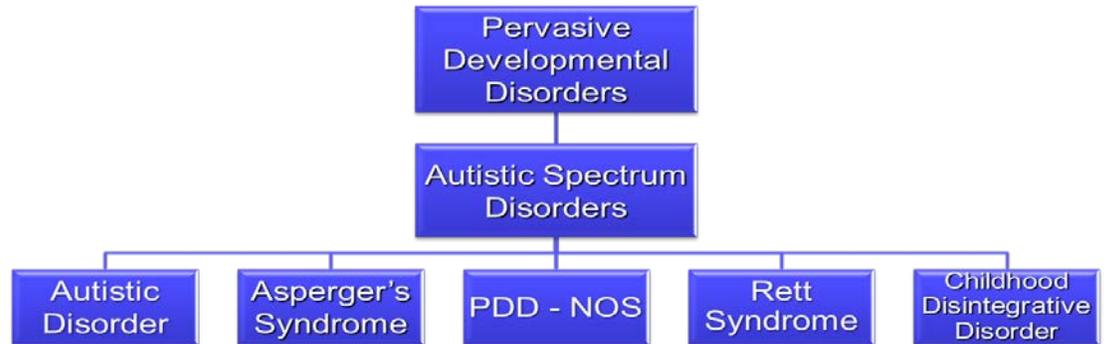


Figure 2 – Proposed DSM-5 Classification for ASD and Social Communication Disorder

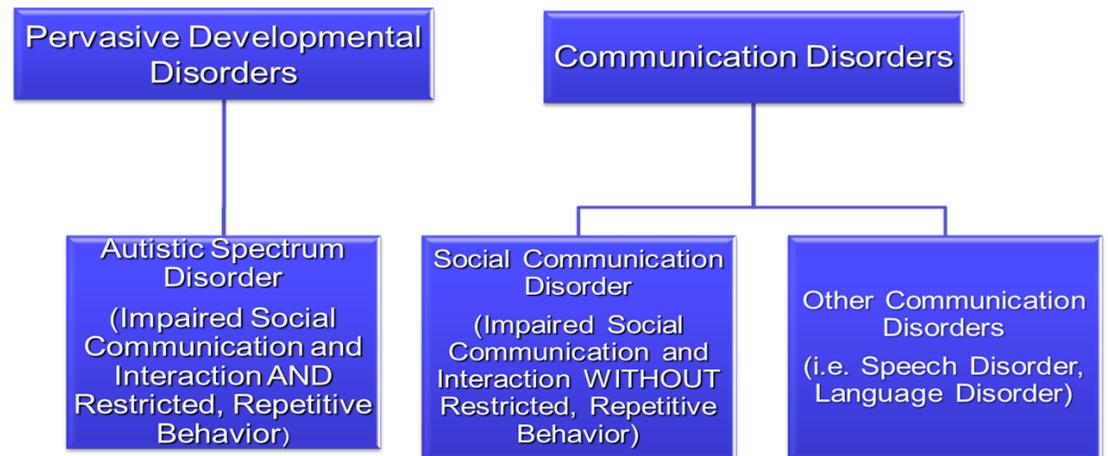
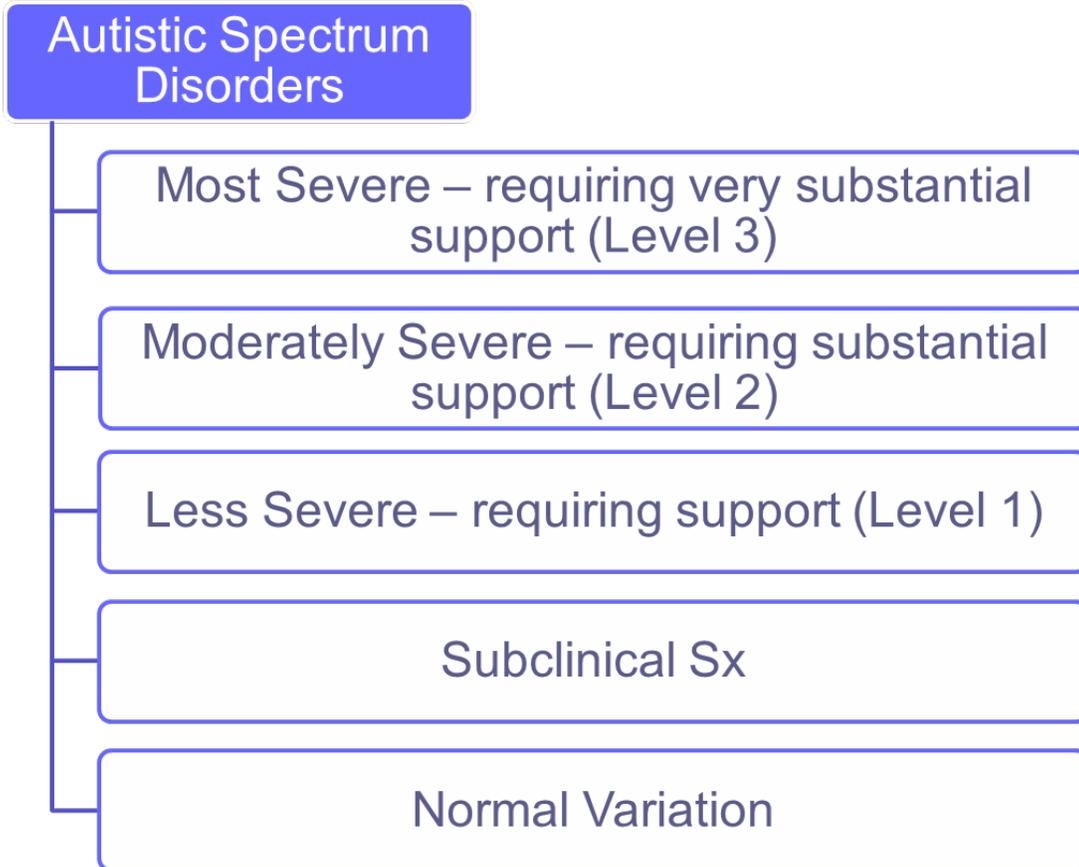


Figure 3 – Proposed DSM-5 Classification of ASDs by severity





Want to learn more about reporting child abuse and neglect?

http://ems.dhs.lacounty.gov/EmergiPress/NewsLetters/EmergiPress2011_2.pdf

UCLA Autism Study

The UCLA Center for Autism Research and Treatment (CART) recently received funding from the NIH to conduct a longitudinal research study of infants at very high-risk of developing autism spectrum disorders (ASD). Infants with a family history of autism have a recurrence rate of 19% to 45%, depending on the number of affected relatives. Infants at very high-risk for autism are children who have more than one sibling with ASD, or one sibling as well as another close relative (parent, aunt, uncle, first cousin) with ASD (or symptoms of an ASD). These infants are nearly 50 times more likely to develop an ASD than infants born in a family without a history of autism.

UCLA is asking for your help in identifying families in your practice who may qualify for participation in the study.

Specifically, informing women who are expecting a high-risk baby about this exciting new study which aims to (1) lower the age at which autism can be diagnosed by identifying early markers of autism before language and behavioral impairments are typically observed, and (2) identify genetic influences and neural mechanisms associated with atypical developmental trajectories. Infants must be under six-weeks of age at the start of the study and will undergo evaluations at 6-weeks, 3, 6, 9 and 12 months (testing will include functional and structural MRI, EEG, eye-tracking, cognitive and behavioral assessments and genetic testing). Ultimately, UCLA hopes to use the knowledge obtained from this study to identify children who may benefit from early intervention and to design treatments that target specific brain dysfunction. Importantly, participating infants who are identified as showing early signs of autism at twelve-months of age will be referred for potential participation in an intervention study led by Dr. Connie Kasari – a leader in the field – which involves a high intensity behavioral treatment specifically designed for young children at high risk for autism.

There is no cost to participate in this study and medical insurance is not required. Interested parents may contact the study coordinator directly at 310-794-2215 or info@autism.ucla.edu. You can view a flyer for parents containing all relevant study information [here](#).

Please contact Susan Bookheimer, PhD if you have any questions. You may reach her at sbook@ucla.edu.

Susan Bookheimer, PhD

Joaquin Fuster Professor of Cognitive Neurosciences
Department of Psychiatry and Biobehavioral Sciences
(310) 794-6386

Are you pregnant? Worried that your baby may develop autism?

If you have a family history of autism,
you may be eligible to participate
in a study of infant development at UCLA

Participating families will receive:

- ◆ \$30-50 for each visit to UCLA
- ◆ Developmental feedback
- ◆ A picture of your baby's brain
- ◆ Free intervention if eligible



Eligible infants are under six weeks old and have either:

- ◆ More than one sibling with autism, or
- ◆ One sibling with autism and an extended family history of the disorder

This study involves several visits to UCLA in your baby's first year and will include:

- ◆ EEG, MRI, and Eye-tracking
- ◆ Developmental assessments



Please contact us for more information:
(310) 825-3478 | siblings@autism.ucla.edu

Thinking Outside of Our Borders:

Global Immunization Advocacy

Gitanjali (Tanya) Arora, MD, MHS, DTMH, FAAP

Pediatric Hospitalist, UCLA Mattel Children's Hospital
Faculty, UCLA Program in Global Health

Every 20 seconds, one child dies from a disease entirely preventable by vaccines. Globally, one in five children lacks access to life-saving childhood vaccinations. I have always had an interest in vaccine preventable diseases. My earliest childhood memory is of being in a hospital waiting room as my infant brother struggled with respiratory syncytial virus (RSV). I desperately wanted him to be well, and when the pediatrician came to tell us that my brother would be ok, I knew I wanted to be just like this doctor someday. My desire to care for children was reinforced by summer vacations in India, seeing kids who looked just like me by the roadside unable to walk or play because they had been afflicted by polio. At that age, it was difficult for me to understand the inequity that existed in my two home countries. My father grew up in India and contracted small pox as a child. He explained that this was a terrible disease and he was lucky to be alive, although he would carry the stigma of pox virus scars on his face for life. From early on, I learned how lucky I was to grow up in a time and country when my parents did not have to worry about life-threatening illnesses each time I had a fever. I also learned that it was just a twist of fate that separated my life from the lives of the children by the roadside. I realized I had a responsibility to use my skills and the opportunities I had been given to help children, regardless of where they lived.

Shortly after residency, I signed up to work internationally with Doctors Without Borders in South Sudan. Here, I found myself caring for children with diseases that were entirely preventable. Tetanus, polio, and meningitis—diseases my mentors and I had never seen before—were common occurrences. Vaccines in South Sudan are only made available to children less than one year old and because of the poor health care infrastructure, most children remain unvaccinated. In our 52-bed unit hospital, I had nine beds that were always full of infants and children with tetanus. They would spasm with the slightest sound or touch. Two afternoons a week, when an airplane bringing much-needed medical supplies would land at the field nearby, all nine children would go in to simultaneous spasm. The other beds were filled with patients with diarrhea, pneumonia, pertussis, and a few with meningitis. Most of the hospitalizations and subsequent deaths could have been prevented with vaccines. Given the tenuous political state of South Sudan, it was clear that if our hospital did not exist, children with diseases such as diabetes, tuberculosis, and severe burn injuries would certainly die. Nearly two-thirds of my pediatrics ward was filled with children whose illnesses could have easily been avoided. If they had access to vaccines, they would not have become sick and would not need hospitalization. I cannot state strongly enough the impact that vaccination would have in this unstable community.

By the time I left South Sudan, I had become sickened with myself for having said to parents, "There is nothing I can do" for their child's breathing problem, brain infection, seizures, dehydration, and spasms for tetanus. As a matter of fact, something could have been done. These children should have been vaccinated. The inequity that exists in the fact that more than one out of four children in so many places in the world will not make it to their 5th birthday is irreconcilable, especially knowing that in my hometown of Los Angeles, parents feel comfortable refusing life-saving vaccines for their children because of the safety and comfort we live in.

Recently, the American Academy of Pediatrics (AAP) and the United Nation Foundation's Shot@Life Campaign offered me the chance to share my experiences with senators and representatives here in Los Angeles. The Shot@Life campaign empowers Americans to champion vaccines as a cost-effective way to save lives of children in developing countries. I was joined by other champions from Shot@Life, including Tracey Clark, a blogger whose globally-minded daughter called her to action to be an advocate for all children regardless of where they lived, and Amanda Peet, an actress, who learned about the importance of vaccines while caring for her own children. Through Shot@Life, I have been inspired by pediatricians, including Heide Woo and Natasha Burgert, who not only protect their own patients but also inform parents about the importance of vaccinations for children beyond the borders of their office. Natasha created an informative video that can be seen at <http://kckidsdoc.com/i-hate-measles-a-summer-project-for-shotlife.html>. For now, the federal government is operating under a continuing resolution that will keep initiatives, such as the CDC global vaccination program, operating until March 2013. When Congress revisits the budget process in 2013, the AAP and Shot@Life will continue to be a voice for children's health, both here and abroad.

The story of my brother with RSV ends well. He is now a healthy adult and is expecting his own child this spring. He plans to give the best care possible for his child, ensuring full vaccination in accordance with the AAP-recommended immunization schedule. We feel tremendously fortunate to have access to these life-saving vaccines. To help all parents care for their children, pediatricians can support global vaccination and eradication efforts by continuing to assure high levels of immunization coverage in their patients and by partnering with others in their community such as Rotary Clubs and the United Nation Foundation's Shot@Life Campaign to help promote global vaccination efforts.

For more information, visit the AAP global immunization webpage:
<http://www2.aap.org/international/immunization/default.html>

ON THE
WEB



For more
information on RSV
and prevention

visit <http://www.cdc.gov/rsv/about/index.html>

Information on flu
vaccines

visit <http://publichealth.lacounty.gov/ip/flu/FluLocatorMain.htm>

ISMP National
Vaccine Error
Reporting
Program

<http://verp.ismp.org/>



Vascular Birthmarks on the Face:

How to Distinguish among a Hemangioma, Port-Wine Stain, and Nevus Simplex

Ki-Young Suh, MD

Director of Pediatric Dermatology, Assistant Clinical Professor
Division of Dermatology, David Geffen School of Medicine at UCLA

The term “vascular birthmark” can refer to several different vascular malformations and growths that present on the skin of a newborn at birth or shortly thereafter. The three common ones are the hemangioma, port wine stain, and nevus simplex (also known as salmon patch or angel’s kiss). Distinguishing among them may not be difficult in their classic form. However, at times their features may overlap, making the diagnosis less clear. This article will discuss 1) how the features of these three common vascular birthmarks may overlap, especially when they present on the face, and 2) why making the correct diagnosis is important.

Hemangiomas are the most common benign tumors of infancy, and they usually display a unique and characteristic growth history that greatly aids in their recognition. Hemangiomas can be present at birth as a flat patch of telangiectasias (dilated capillaries), a bruise-like patch, or an area of pallor. Shortly after birth the growth phase begins, and the hemangioma may rapidly increase in size until about 3-5 months of age. The growth phase then slows, after which the size of the hemangioma stabilizes, and then the hemangioma slowly involutes over the ensuing years¹.



Figure 1 - Reprinted with permission.²



Figure 2 - Reprinted with permission.³

An important exception to this rule is the small subset of hemangiomas that have absent or minimal growth phase. They look like early or precursor hemangiomas and can present as a pink, faint telangiectatic patch; a darker red, coarsely telangiectatic patch; or a bruise-like patch (figures 1, 2). These hemangiomas may be associated with a small component of growth, but overall are arrested in their early growth phase².

Port wine stains (PWSs) fall under the category of vascular malformations rather than tumors, and they are more specifically capillary malformations. They are present fully formed at birth, and they do not undergo regression. The color may vary from pink to purplish-red, and in general, the PWS has an even, homogeneous appearance (figure 3). On close inspection, fine telangiectasias may be appreciated, but it is not typically characterized by coarse telangiectasias. The natural history of PWSs is that they will likely darken over years, and a portion of them may be associated soft tissue hypertrophy and swelling⁴.



Figure 3.

Figure 3

Nevus simplex is a common vascular birthmark that presents as a pink-to-red patch on the mid-forehead and/or the upper eyelids, nose and philtrum. They often fade without treatment within the first or second year of life, although it is not uncommon for them to fade partially and persist into adulthood⁵.

As can be seen in Figures 1 -3, the features of minimally proliferative hemangiomas, PWS, and nevus simplex can be deceptively similar. It is important to identify these entities correctly for several reasons. First, as mentioned above, the natural history is different. Whereas hemangiomas and nevus simplex will involute and fade on their own over several years, PWS will remain fixed and may even darken and become thicker over time. The second reason relates to treatment considerations. Whereas minimally proliferative hemangiomas and nevus simplex will fade without treatment, PWS will require laser treatment in order to help lighten the red-to-purplish color. Even after good results are obtained with laser treatment, over time the PWS has been observed to darken again, and patients may wish to seek repeat treatments.

The third reason deals with the associated disorders that may exist with these vascular lesions:

- *PHACE syndrome*. Hemangiomas that cover a broad surface of the skin and appear territorial rather than localized, are called segmental. Segmental hemangiomas on the face are associated with structural defects in a syndrome known as PHACE syndrome (posterior fossa malformation, large facial hemangiomas, arterial anomalies, cardiac anomalies, and eye anomalies). In order to evaluate for these abnormalities, an MRI and MRA with contrast, cardiac echocardiogram, and referral to an ophthalmologist may be necessary.⁶
- *Hemangiomas* in the beard area and airway involvement. Hemangiomas that are present in the "beard area," such as the preauricular area, mandible, chin and lips, can be associated with hemangiomas in the upper airway. Even when the hemangioma on the face is minimally proliferative, the airway component may undergo a rapid and significant growth and result in airway obstruction and breathing distress.⁷
- *PWS* on the face can be associated with Sturge-Weber syndrome, which is characterized by vascular malformations in the leptomeninges and the eye. Sturge-Weber syndrome is a risk when the PWS is present along the first branch of the trigeminal nerve (V1), with a risk of about 8%. If the PWS extends more broadly into V2/V3 or affects V1 bilaterally, the risk is higher at approximately 25%.⁸

Because of the important associated disorders that require careful evaluation and management, referral to a pediatric dermatologist is strongly recommended when the diagnosis of a vascular birthmark is not clear.

The following is a table to summarize and highlight important points:

	Clinical Appearance	Natural History	Potential Associated Disorders	Management
Minimally proliferative hemangiomas	1. Pink or red telangiectatic patch with or without surrounding pallor; the telangiectasias tend to be more uneven, with fine and coarse components AND/OR 2. Bruise-like patch	They lack the typical, rapidly proliferative growth phase and involute slowly over several years.	1. PHACE syndrome when a large hemangioma is present on the face: posterior fossa malformation, arterial anomalies, cardiac anomalies, eye anomalies 2. Airway hemangioma when hemangioma is in "beard area"	1. MRI/MRA with contrast, cardiac echo, referral to ophthalmology 2. If stridor or airway distress is present, then refer to otolaryngology to evaluate for airway hemangioma
Nevus simplex	Pink or red telangiectatic patch on the mid-forehead, upper eyelids, nose, or philtrum	Fades spontaneously within 1-2 years of life.	--	--
Port wine stains	Homogeneous pink, red, or red-purple patch	Fixed. May darken and even thicken over many years.	When present on V1 +/- V2 or V3, Sturge Weber syndrome	Consider MRI based on presence of CNS symptoms (e.g., seizures, hemiplegia, mental retardation) Referral to ophthalmology

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