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EHDI *Express*

Early Hearing Detection & Intervention
a program of the American Academy of Pediatrics

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Front Page Feature

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How Many Baseball Caps Do You Wear?

A baseball cap, literally and figuratively, is important to help one focus on the task at hand.

For instance, long ago my uncle who was convinced that work is for folks who do not know how to fish, showed me that fish would not bite unless the angler literally wore a well-set baseball cap with politically incorrect writing. From my day job, I knew that wearing a baseball cap would not only keep the sunlight from blazing into my eyes but would also help me to get where I wanted to go! Yes, the baseball cap disturbs the coiffure, but being able to travel easily is better.

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Otherwise said, a figurative baseball cap helps me to narrow my scope of work—of what's next. In my clinical care of children, that task is working to define for each individual patient the ear "problem," what that problem amounts to, checking that the data "adds up," (Audiologists, remember the "cross-check principle" that James Jerger, PhD and Linda Hood, PhD so well espoused a couple of generations ago—"No result should be accepted until it is confirmed by an independent measure."?) what course of treatment to recommend, and then executing the plan.

When I worked for 10 years in the United States Public Health Service Indian Health Service, my figurative baseball cap was invaluable in helping me to attend to the individual patients, especially during our "reservation clinics." Away from the monitoring eyes of the bureaucracy, it was during these clinics that we could truly focus on individual patient needs—not worrying so much about the official top-down edicts. We kept our souls clean.

When working with these patients in the Indian Health Service, especially those with hearing problems, who clanged and clawed for every decibel to communicate, I came to appreciate the need for yet another baseball cap (at least figuratively). I needed a baseball cap to help me think beyond the individual, to think about the population in which that individual existed. I came to realize the need to think more broadly in terms of a population's hearing resources and equipment and time commitment, so that each patient may be able to communicate to her/his maximum potential.

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The follow-up from an individual newborn's auditory physiologic screening, whether automated OAE or A-ABR, must be logged to the county, region or state, so that we (the big picture royal "we") can know that resources are being wisely used; so that we know when we can celebrate what is working; so that we can address what is not working so well, and we can aim to help cultivate a lifetime of better communication. That is, we as providers and EDHI screeners should wear two baseball caps—one to remind us to acquire the data and communicate the result, (did they pass THIS SCREENING, THIS MORNING; or did not pass THIS SCREENING, THIS MORNING) emphasizing that the checking is NOW and for this modality—not necessarily a harbinger of future doom or gloom; and another for the future, to help us to facilitate "secondary screenings" and/or monitor language development.

In working with audiologists, we should recognize that they too should be wearing caps; the practical baseball cap of thoroughness and the tough baseball cap to help resist the hierarchal-demanded good-looking template-based fast report that can so easily harbor garbled incoherencies.

For you grammarians dismayed by my habitual run-on sentences, please know that my uncle did fish the same reservoir as did William Faulkner. Run-ons are how some of us think—though Faulkner preferred to fish wearing a non-baseball hat.

Clinical Corner

A Small Molecule Mitigates Hearing Loss in a Mouse Model of Usher Syndrome III

A new study published in Nature Chemical Biology reports the first small molecule targeted therapy for progressive hearing loss in a mouse model of USH3, an USH classified by progressive loss of hearing and vision starting in the first few decades of life along with variable balance disorder. This is caused by destabilizing mutations in the gene encoding the clarin-1 protein (CLRN1).

The finding of this targeted therapy in a mouse model holds much promise for the thousands of patients and their families diagnosed with USH3 for whom symptoms can be devastating, and for the physicians who have not yet been able to offer a treatment to preserve hearing or vision in these individuals.

This research was conducted by a team of scientists at Case Western Reserve University School of Medicine who developed a small molecule to mitigate the effects of the common USH3 mutation Clarin-1-N48K (CLRN1N48K). Members of the team included chemists from BioFocus (United Kingdom) who optimized the original 'hit' small molecule identified at the School of Medicine, and university researchers who characterized the biochemical properties of the molecules.

The team utilized a novel research strategy involving a cell-based high-throughput screening of small molecules capable of stabilizing the mutant CLRN1N48K, a secondary screening to eliminate general proteasome inhibitors, and finally an iterative process to optimize structure activity relationships, resulting in the identification of the compound BF844. To test the efficacy of BF844, researchers developed and studied a mouse model that mimicked the progressive hearing loss of USH3, concluding that BF844 effectively attenuated progressive hearing loss and prevented deafness in this model.

Source: Alagramam KN, Gopal SR, Geng R, et al. A small molecule mitigates hearing loss in a mouse model of Usher syndrome III. *Nature Chemical Biology.* 2016.

Quality Improvement (QI) Buzz

The Sum Is Greater Than Its Parts: Synergistic Improvement When EHDI Programs and Chapter Champions Work Together

Below are three examples of how state Early Hearing Detection and Intervention (EHDI) programs have engaged their AAP EHDI Chapter Champion in quality improvement (QI).

Louisiana (LA)

Chapter Champion, Dr Thiravat Choojitarom, is involved with LA EHDI in a variety of ways. Dr Choojitarom serves as the chairperson of the state EHDI Advisory Council, which guides the state EHDI program on issues relevant to QI. He also collaborates with state EHDI staff in research and data collection to help identify needs, and then works with EHDI staff to develop strategies to test strategies that may fill said needs. Dr Choojitarom also serves as a liaison to other primary care providers by providing information and updates about ongoing state EHDI QI work at state and regional AAP chapter meetings. Dr Choojitarom works with LA Hands & Voices chapter to help identify ways physicians can keep families engaged. Having a committed and involved chapter champion greatly extends the reach of the state EHDI program and leads to higher quality outcomes in the LA EHDI program.

Colorado (CO)

Dr Maureen Cunningham, the new CO Chapter Champion, has been eager since day one to become involved with the state's EHDI QI work. While recently completing her Master's in Public Health, Dr Cunningham evaluated all birthing hospitals in CO about their newborn hearing screening processes and procedures. She used the data collected to identify areas of loss to follow-up (LTFU) and in turn worked with EHDI program staff to brainstorm and test strategies to improve LTFU. Dr Cunningham also participated in a QI project that involved outreach to notify physicians when a child in their practice is reported as not passing two hearing screenings. Dr Cunningham is helping physicians become familiar with the new integrated immunization registry data—that now also includes EHDI information and data. The CO EHDI program is grateful to have the help of Dr Cunningham and looks forward to the improved outcomes she hopes to help them reach.

Vermont (VT)

Dr Hassler, the VT EHDI Chapter Champion, is the primary investigator on the VT Centers for Disease Control and Prevention cooperative agreement and Health Resources and Service Administration (HRSA) grant. Each year Dr Hassler reviews all progress reports and applications for federal partners and shares her perspective on VT QI and data initiatives. Dr Hassler is also key in connecting the EHDI program with external stakeholders. For example, in the neuro-medical clinic that oversees all neo-natal intensive care unit (NICU) babies long-term, Dr Hassler was concerned that NICU infants did not have newborn screening/diagnostic information in their charts and therefore these babies counted toward VT LTFU rates. She brought this to the attention of the VT EHDI staff who are now working with the physician who oversees the neuro-medical clinic to remedy the issue. Dr Hassler was integral to the initial design and implementation of the primary care provider (PCP) pilot study, including reviewing goals and objectives and identifying key stakeholders for participation. The PCP pilot sites have been one of the most successful QI initiatives in reducing LTFU in VT. As hearing screenings/re-screenings increased in the PCP offices, LTFU rates decreased. The VT EHDI program works as a highly functioning collaborative team of which Dr Hassler is a respected and valued member.

Medical Home Resources

Thinking Outside the Box: How to Advance Health Equity and Care Quality in the Pediatric Medical Home Webinar Series, May – June 2016

The National Center for Medical Home Implementation (NCMHI) is facilitating a three-part webinar series focused on providing pediatric clinicians, including those who care for children who are deaf or hard of hearing, with tools and strategies to enhance patient and family experience in the pediatric medical home. Upcoming webinars focus on communicating effectively with patients and families as well as understanding and addressing social factors that shape child health. For more information, visit the NCMHI Web site.

And More….

NIDCD Launches Enhanced Website

The National Institute on Deafness and Other Communication Disorders (NIDCD) has implemented several enhancements to the NIDCD website. The changes are designed to enhance users' experience and strengthen the institute's alignment with federal mandates for information technology. Key features of the revised website include the following:

- Responsive design, enabling the content to automatically adjust to fit a user's screen size, whether it is viewed on a computer, tablet, or phone.
- Redesigned research and funding sections, and a new, prominent section highlighting NIDCD-supported research training and career development funding opportunities throughout the United States and at the NIDCD labs and clinic in Bethesda, Maryland.
- Improved home page design, featuring enhanced navigation and the NIDCD Twitter feed.
- Integration of relevant NIDCD-supported research results with consumer health information on hearing, balance, taste, smell, voice, speech, and language disorders.
- New portal to Spanish-language consumer information published by the NIDCD.

Family Partnerships

Family-to-Family Health Information Centers

Family-to-Family Health Information Centers (F2F HICs) are non-profit, family staffed organizations that assist families of children and youth with special health care needs and the professionals who serve them. Pediatric clinicians, family advocates, and practice administration can connect with their F2F HIC to increase the coordination of care and support provided to families. Multiple F2F HICs are involved in medical home implementation projects in their states, and as such, data indicate that F2F HICs are successful in helping families navigate systems, partner with professionals in decision-making, and learn and/or connect with community services.

To reinforce the *partnership* component of your Chapter Champion work plan, find out more about the F2F HIC in your state. Focus on learning more about the activities and resources they offer and determining if there is potential for future collaboration on EHDI-related efforts.

Upcoming Events & Opportunities

Event/Opportunity	Date	More Details
Webinar: Changing Relationships: How to Foster Effective Communication with Patients and Families	May 31, 2016 11am CT	Register
Webinar: Changing Practice: How to Understand and Address Social Factors that Shape Child Health	June 8, 2016 11am CT	Register
53rd Biennial National Association of the Deaf Conference	July 5–9, 2016, Phoenix AZ	Website
Cytomegalovirus Public Health & Policy Conference	September 26-27, 2016, Austin TX	Website
AAP National Conference & Exhibition	October 22-25, 2016 San Francisco CA	Website

The AAP EHDI program implementation staff send this e-mail update to the Academy's EHDI Chapter Champions, other interested AAP members, state EHDI coordinators, and other stakeholders. For additional information on hearing screening and to access previous editions of the EHDI E-mail Express, click here. If you would like to unsubscribe to the update contact Michelle Esquivel at mesquivel@aap.org or 847/434-4989.