

Update on the Iowa Early Hearing Detection and Intervention (EHDI) Program

Newborn Hearing Screening

Congenital hearing loss affects 2 to 3 per 1000 babies born in the United States (1,2). In Iowa, this means that over 80 babies per year are born deaf or hard-of-hearing. Because undetected prelingual hearing loss results in childhood speech and language delay and because early identification and intervention vastly improve outcomes for children, 42 states now have legislation that mandates universal newborn hearing

screening. Only 6 of these require written consent of parents to perform the screen, which demonstrates the establishment of newborn hearing screening as a standard of newborn care. Objective, physiological hearing screening during the birth admission has been endorsed by the American Academy of Pediatrics, the Joint Committee on Infant Hearing and government health agencies. Prior to the implementation of newborn hearing screening, the average age of identifica-

continued on page 6

EDITOR: Stephen K. Hunter, M.D., Ph.D., Obstetrics and Gynecology; stephen-hunter@uiowa.edu. ASSOCIATE EDITOR: Michael J. Acarregui, M.D., Associate Professor, Pediatrics; michael-acarregui@uiowa.edu. EDITORIAL ASSISTANT: Kathy Brogden, Pediatrics; katherine-brogden@uiowa.edu. Call 319-356-2637 to be added to the mailing list.

Iowa City, IA 52242-1083
200 Hawkins Drive
Department of Pediatrics
Statewide Perinatal Care Program

The Iowa
PERINATAL
Letter

Update on the Iowa Early Hearing Detection and Intervention (EHDI) Program

continued from page 5

tion of congenital hearing loss in the United States was 2 ½ years (3). This age of identification has dropped dramatically since the achievement of nearly universal newborn hearing screening (4). Along with technological advances in hearing aids and cochlear implants, the result has been exciting progress in speech, language, academic, social and emotional outcomes for children who are deaf or hard-of-hearing and their families.

The Iowa EHDI program has benefited from continuous federal funding from the Centers for Disease Control and Prevention and the Bureau of Maternal and Child Health. For many years, Iowa had a high rate of voluntary screening and Iowa EHDI legislation went into effect on January 1, 2004. Thanks to the efforts of many stakeholders in our hospitals, Area Education Agencies (AEAs) and state agencies, the Iowa EHDI program can celebrate much success. By 2007, 99% of all newborns in Iowa had their hearing screened by objective methods during the birth admission.

Monitoring Infants at Risk for Delayed Onset Hearing Loss

Consensus statements about best practices in EHDI programs have been periodically published by the Joint Committee on Infant Hearing (JCIH) since the 1960's. Joint Committee member organizations include the American Academy of Audiology, the American Academy of Pediatrics, the American Academy of Otolaryngology-Head and Neck Surgery, the American Speech-Language-Hearing Association, and the Council on Education of the Deaf. In their most recent statement published in the Fall of 2007 (5), the committee outlined revised and much improved recommendations for monitoring babies who may pass the newborn hearing screen, but are at risk for developing delayed onset hearing loss that can also adversely impact speech and language development and social and emotional growth.

There are some health conditions or medical history factors that put a baby at particular risk for delayed onset hearing loss. Newborn hearing screening will detect congenital hearing loss, but by age 4, prevalence of permanent childhood hearing loss will grow by 45% (6). These babies will often pass the newborn hearing screen, but develop significant permanent hearing loss in the first few months or years of life. Parents and primary care providers may even disregard their own

concerns about hearing because the baby passed the newborn screen. The previous recommendation of the JCIH, published in 2000 recommended audiological evaluation of babies with any of these risk factors every 6 months. It was quickly recognized that this approach was neither feasible nor cost-effective. In their 2007 position statement, the committee recommended that these risk factors be tracked and that babies at high risk for developing delayed-onset hearing loss receive an audiological evaluation, with the recommended schedule based on the particular risk factor. A recent change in Iowa EHDI legislative rules, signed by the governor in April 2009, requires reporting of these known risk factors into the Iowa EHDI database.

Risk factors for delayed-onset hearing loss that were designated by the JCIH are listed in Table 1. Several of these are actually postnatal risk factors, but the committee included them as a guide for primary care providers to help decide what children need audiological referral, along with on-going medical and speech-language surveillance. The committee recommends that babies with risk factors should be seen by an audiologist by 6 months or at least once by 24 to 30 months of age (depending on the risk factor) and, of course, sooner if hearing loss is suspected or speech or language milestones are delayed. There is some misconception that an audiological referral will always result in an auditory brainstem response (ABR) test and the cost might not be justified. This is not true. Although an audiological assessment of a young child sometimes requires an ABR, audiologists can use other physiological tests and behavioral methods to assess a young child's hearing sensitivity.

Children who have risk factors that are highly associated with delayed-onset hearing loss should receive more frequent audiological assessments. Those risk factors that are usually known at birth and are highly associated with delayed onset hearing loss are family history of permanent childhood hearing loss, extracorporeal membrane oxygenation (ECMO), confirmed congenital cytomegalovirus infection (CMV) and syndromes associated with late onset hearing loss. There is a strong relationship between ECMO treatment and sensorineural hearing loss in children, with 7.5% of ECMO survivors experiencing sensorineural hearing loss (7). This is significantly higher than the 1-3% of the general NICU population. Hypoxia or the use of associated ototoxic medications are more likely the cause of hearing loss than ECMO treatment itself. However, the strong correlation has caused the JCIH to recommend that ECMO survivors receive close monitoring of speech, language and hearing milestones. Congenital CMV is also strongly associated with late-onset hearing loss: In symptomatic newborns, about 16% will have congenital hearing loss with 36% having

hearing loss by 6 years of age. In asymptomatic newborns, 3% will be born with hearing loss and 11% will have permanent hearing loss by age 6 (8).

The Iowa EHDI Program for Surveillance of Children with Risk Factors for Delayed-Onset Hearing Loss

The Iowa Early Hearing Detection and Intervention (EHDI) program recently began sending letters to families and primary healthcare providers (PCPs) of children with risk factors associated with permanent hearing loss. The letters provide information about

recommended follow-up and resources for more information. We need your help to make this process efficient and accurate. Participants in newborn hearing screening and follow up can help with this effort in many ways: Please notify families the child has a risk factor and explain that a hearing assessment will be needed at a later date. Please mark risk factors in eSP (the Iowa EHDI database) records. Iowa EHDI can not notify families and PCPs if they do not know about the risk factors. Please make sure the risk factors are correct and confirmed. Please list the PCP who will see the child after discharge from the hospital.

Table 1: JCIH (2007) Risk factors associated with permanent childhood hearing loss (* indicates greater concern for delayed onset hearing loss)

- Caregiver concern* re hearing, speech, language or developmental delay
- Family history* of permanent childhood hearing loss
- NICU stay of >5 days, which may include ECMO* assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/lasix) and hyperbilirubinemia requiring exchange transfusion
- Confirmed in-utero infections, such as CMV*, herpes, rubella, syphilis and toxoplasmosis
- Craniofacial anomalies, including those involving the pinna, ear canal, ear tags, ear pits and temporal bone anomalies
- Syndromes associated with hearing loss or progressive or late onset hearing loss* such as neurofibromatosis, osteopetrosis and Usher's syndrome. Other frequent syndromes include Waardenburg, Alport, Pendred and Jervell & Lange-Nielson
- Neurodegenerative disorders* such as Hunter syndrome or sensory motor neuropathies, such as Friedrich's ataxia and Charcot-Marie-Tooth syndrome.
- Culture-positive postnatal infections associated with sensorineural hearing loss* including bacterial and viral (esp. herpes and varicella) meningitis
- Head trauma esp. basal skull/temporal bone fracture*
- Chemotherapy*

—Lenore Holte, Ph.D., CCC-A
 Clinical Professor
 Dept. of Speech Pathology and Audiology
 Dept. of Pediatrics
 University of Iowa

References

1. Directors of Speech and Hearing Programs in State Health and Welfare Agencies (DSHPSHWA, 2003). Infants tested for hearing loss, United States, 1999-2001. In *Morbidity and Mortality Weekly Report*, Oct 17, 2003, 981-984.
2. White, K (2003) The current status of EHDI programs in the United States. *Mental Retardation & Developmental Disability Research Reviews*, 9 (2), 79-68.
3. Harrison M, Roush J (1996). Age of suspicion, identification and intervention for infants and young children with hearing loss: A national study. *Ear and Hearing*, 17, 55-62.
4. Vohr BR, Carty LM, Moore PE et al. (1998). The Rhode Island Hearing Assessment Program: Experience with statewide hearing screening. *Journal of Pediatrics*, 133, 353-357.
5. Joint Committee on Infant Hearing (2007). Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics*, 120; 898-921.
6. Morton CC, Nance WE (2006). Newborn hearing screening a silent revolution. *New England Journal of Medicine*, 354 (20), 2151-2164.
7. Fligor B (2008). Hearing outcomes in the most critically ill neonate population. *Audiology Today*, 20, 8-16.
8. Dahle AJ, Fowler KB, Wright JD, Boppana SB, Britt WJ, Pass RF (2000). Longitudinal investigation of hearing disorders in children with congenital cytomegalovirus. *The Journal of the American Academy of Audiology*, 11 (5), 283-290.

Neonatal Circumcision: Educating Parents

According to the National Hospital Discharge Survey, the overall circumcision rate in the United States in 2006 was 59.1%. Rates were highest in the North Central Region, which includes Iowa, at 77.9%. Despite being a rather common procedure, The American Academy of Pediatrics does not recommend routine neonatal circumcision. However, parents should be provided with accurate and unbiased information in order to determine if circumcision is in the best interest of their child. There have been numerous studies published regarding possible medical benefits related to neonatal circumcision. Evidence suggests a decreased risk of urinary tract infections in the first year of life, as well as a decreased risk of penile cancer and transmission of sexually transmitted diseases in adulthood. However, the low incidence of urinary tract infections and penile cancer in uncircumcised males and the impact of condom use on decreasing the risk of STD transmission make mandating neonatal circumcision difficult.

If families elect to proceed with circumcision, we must inform them the procedure comes with risk, particularly bleeding, infection, and poor cosmetic outcome. Certainly, one of the most common reasons for a circumcision revision is cosmetic appearance. Parents should be educated on the subjective nature of the procedure, particularly regarding the amount of foreskin to remove. It can be difficult to estimate the appropriate amount of tissue to remove on an infant penis, because the penis will change considerably as the child ages. This means that small differences in foreskin removal at the time of circumcision may make big differences in the circumcised penis in adulthood. A consequence of taking off too much skin can be pain or skin tearing with erection. Due to this, most clinicians who perform circumcision tend to be more conservative when deciding how much foreskin to remove.

Parents should be educated on the typical appearance and proper care of the circumcised penis. A prospective study in 1997 by Van Howe evaluated the different clinical appearances of the circumcised penis. This study revealed a wide variability in the appearance of a penis following circumcision. Circumcised males under the age of 3 years of age were significantly more likely to have a partially or completely covered glans. However, circumcised males over the age of 3 years were more likely to have a fully exposed glans, particularly after reaching Tanner Stage V. These findings suggest that circumcised males with redundant foreskin often "grow into" their circumcision, and as physicians we should counsel parents against costly and unnecessary circumcision revision for cosmetic concerns.

According to Van Howe, circumcised males less than the age of 3 years were also more likely to have a non-cosmetic penile problem such as adhesions, skin bridges, trapped epithelial debris, meatitis, preputial stenosis, or balanitis when compared to their uncircumcised peers. Since many of these problems are related to hygiene we should counsel parents on

the proper care of a circumcised penis. A circumcised male needs to have the skin overlying the glans pulled back and cleaned regularly until the age of 15-18 months. This practice will assist in preventing the formation of adhesions, skin bridges, and debris accumulation.

In regards to adhesions, there is some controversy in terms of their management. Certainly, circumcised males with any skin overlying the glans are more likely to form adhesions, which emphasizes the importance of good hygiene. The mean age for developing adhesions is 6-8 months of age. Some urologists will manually take down adhesions, while others feel adhesions can be observed without any intervention, noting that the incidence of adhesions decreases with increasing age. Hypotheses for the natural decrease in adhesions with age include nocturnal erections, penile growth, and hormonal influences. Another treatment option for penile adhesions is topical betamethasone cream, which has been used as medical management of phimosis in uncircumcised males for many years. The application of betamethasone cream, 0.05-0.1%, 2-3 times daily for 3-4 weeks with gentle retraction by the parents at home is the approach many general pediatricians recommend. Palmer et al. published a small study in 2005 which showed a success rate of 79% with the use of betamethasone cream as treatment for cicatricial scars following circumcision.

In closing, circumcision is an elective procedure with definite risks and complications. It is not a procedure to be taken lightly and our patients' families need to be properly educated about it. As physicians it is also important for us to recognize normal variants in the circumcised penis, educate our families on its proper care, and treat common problems in order to prevent unnecessary referrals and revisions.

—Lisa Didion, MD
Fellow Associate
University of Iowa Children's Hospital
Division of General Pediatrics
and Adolescent Medicine

References:

- American Academy of Pediatrics, Task Force on Circumcision, Circumcision Policy Statement. *Pediatrics*. 103 (3): 686-693, March 1999. Reaffirmed May 2005.
 - Palmer JS, Elder JS, Palmer LS. The use of betamethasone to manage the trapped penis following neonatal circumcision. *J Urology*. 174: 1577-8, Oct 2005.
 - Ponsky LE, Ross HR, Knipper N, Kay R. Penile adhesions after neonatal circumcision. *J Urology*. 164: 495-496, Aug 2000.
 - Van Howe RS. Variability in penile appearance and penile findings: a prospective study. *Brit J Urology*. 80: 776-782, 1997.
- The Circumcision Reference Library



This letter is funded by a community award from the March of Dimes Iowa Chapter