### Policy
Children with risk factors related to late onset or progressive hearing loss will be monitored so that if hearing loss develops, it will be detected as early as possible.

### Background
The Iowa EHDI High-Risk Monitoring Protocol is based on the Joint Committee on Infant Hearing 2007 position statement. Emphasis is placed on follow-up as deemed appropriate by the primary health care provider and audiologist. The Iowa protocol describes the follow-up process for children with risk factors.

### Hospital/AEA/Private Audiologists/Health Care Provider Protocol
Best practice indicates that risk factors for late onset hearing loss be reported through the statewide EHDI data management or by reporting the risk factors on the screening and diagnostic reporting forms. Follow-up with families of children at risk for hearing loss can only be successful if risk factors for late onset hearing loss are reported to the EHDI program.

See the appendix for a list of risk factors for late onset hearing loss.

### Risk Factors Follow-Up Procedures
These procedures will apply to babies born in 2008 and beyond.

- Each month, EHDI staff members will search for babies born two months prior who have risk factors listed in the statewide EHDI data management system. (For example, in March, staff members will run a report of babies born in January who have a risk factor for hearing loss.)
- Babies will be sorted based on the risk factor they have. The risk factors will determine when follow-up is recommended.
- EHDI staff members will send the letters in Appendix K to families and primary health care providers of children with risk factors. The letters state:
  - The child should see an audiologist for a hearing evaluation by six months of age if one or more of the following risk factors are present:
    - Bacterial and viral meningitis
    - Congenital Cytomegalovirus (CMV) confirmed in infant
    - Extra-corporeal membrane oxygenation (ECMO)
    - Family history of permanent, sensorineural hearing loss during childhood
Monitoring Children with Risk Factors

- Head injury
- Neurodegenerative disorder
- Other postnatal infections
- Parental concern regarding hearing status
- Syndromes
  - The child should see an audiologist for a hearing evaluation by 24 to 30 months of age if one or more of the following risk factors are present:
    - Cranio-facial anomalies
    - Exchange transfusion for elevated bilirubin
    - Herpes infection confirmed in infant
    - NICU stay longer than five days
    - Other congenital infection
    - Ototoxic medications administered
    - PPHN (persistent pulmonary hypertension) associated with mechanical ventilation
    - Rubella infection confirmed in infant
    - Syphilis infection confirmed in infant
    - Toxoplasmosis infection confirmed in infant

- Babies having a risk factor(s) requiring follow-up at six months and a risk factor(s) requiring follow-up at 24 to 30 months will receive only a six month letter.
- The EHDI staff will periodically search for babies born in earlier months so children whose records are entered into statewide EHDI data management system late will receive risk factor follow-up.
Risk Factors Associated with Childhood Hearing Loss

Care Giver Concern of Hearing Loss

Chemotherapy

Congenital Infection - confirmed in infant
Includes: cytomegalovirus, rubella, syphilis, herpes, toxoplasmosis

Craniofacial Anomalies
Includes: Cleft lip or palate, microtia (abnormally small ear), atresia (blocked or abnormally small ear canal), choanal atresia

Family History
A family history of permanent, sensorineural hearing loss during childhood

Head Trauma
Especially basal skull/temporal bone fracture requiring hospitalization

Hyperbilirubinemia
Requiring exchange transfusion

Mechanical Ventilation or Extracorporeal Membrane Oxygenation (ECMO)
Associated with respiratory complications such as bronchopulmonary dysplasia (BPD), persistent pulmonary hypertension of the newborn (PPHN), and respiratory distress syndrome (RDS)

Neonatal Intensive Care Unit (NICU) Admission Greater Than Five Days
Generally indicates health complications, and possible ototoxic medication treatment

Neurodegenerative Disorders
Hunter syndrome, Friedreich’s ataxia, Charcot-Marie-Tooth syndrome

Neurological Conditions and Syndromes
Includes: seizures, hydrocephalus, intra-ventricular hemorrhage, retinopathy of prematurity

Ototoxic Medications
Includes: Gentamycin, Vacomycin, Kanamycin, Streptomycin, Tobramycin

Physical Findings Associated with Sensorineural or Permanent Conductive Hearing Loss
Example: White forelock

Updated 12-22-08
Monitoring Children with Risk Factors

Postnatal Infection
Includes: Bacterial meningitis, herpes, varicella

Recurrent or Persistent Otitis Media with Effusion Greater Than Three Months

Syndromes Associated with Sensorineural, Conductive, or Progressive Hearing Loss
Includes: Trisomy 21-Down syndrome, Goldenhar, Pierre Robin, CHARGE association, Rubinstein-Taybi, Stickler, Usher, osteopetrosis, Neurofibromatosis type II, Treacher Collins

This list of risk factors above is based on the Joint Committee on Infant Hearing Recommendations (JCIH) published Fall 2007.

Sources: