Recommendations for monitoring hearing in children using a risk factor registry

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Background

- To provide recommendations for risk factor registries incorporated within targeted surveillance programs


Queensland’s risk factor registry

- Family history of permanent childhood hearing loss (mother/father/siblings of baby only excluding grommets/ear infections/trauma)
- **Syndromes** associated with hearing loss (e.g., Down Syndrome, FAS)
- Prolonged ventilation = 5 days (IPPV/CPAP)
- Bacterial meningitis (confirmed/suspected)
- Low birth weight = 1500 grams
- Severe asphyxia at birth (convulsions/HIE/PPHN)
- Craniofacial anomalies, e.g., cleft palate (excluding cleft lip & skin tags)
- Hyperbilirubinemia levels = 450µmol/l (Term) or = 340µmol/l (preterm)
- Proven/suspected congenital infection of the baby (Toxoplasmosis, Rubella, CMV, Herpes, Syphilis)
- Professional concern
<table>
<thead>
<tr>
<th>Grade</th>
<th>Recommendation</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Monitor</td>
<td>Collective evidence generally offers strong support for monitoring. e.g., existence of cohort studies indicating cases of postnatal hearing loss in children with the risk factor in isolation + a positive yield + positive relationship/significant Chi-squared correlation + OR &gt; 1</td>
</tr>
<tr>
<td>B</td>
<td>Potentially Monitor</td>
<td>Overall findings are mixed; however, some or most indicate support for monitoring as per grading A</td>
</tr>
<tr>
<td>C</td>
<td>Lack of Evidence</td>
<td>Collective evidence is lacking. e.g., no literature evidence or case studies only; +/- presence of complicating risk factors; and/or logistic regression not completed. Alternatively, overall findings may be highly mixed/inconclusive.</td>
</tr>
<tr>
<td>D</td>
<td>Potentially Don’t Monitor</td>
<td>Overall findings are mixed; however, some or most indicate support for not monitoring as per grading E</td>
</tr>
<tr>
<td>E</td>
<td>Don’t Monitor</td>
<td>Collective evidence generally offers strong support for not monitoring. e.g., existence of cohort studies indicating no/limited cases of postnatal hearing loss and complicating risk factors present + nil yield + negative relationship/insignificant Chi-squared correlation + OR = 1</td>
</tr>
</tbody>
</table>
Family History

- Limited evidence

Three studies were identified that reported on family history and postnatal hearing loss (Robertson et al., 2009; Thiringer et al., 1984; Weichbold et al., 2006)

- Difficult to establish the nature of the relationship as (i) children with a hearing loss only were included; or (ii) the children who developed a postnatal hearing loss had more than one risk factor

Support in favour of monitoring

- Family history present in 46.4% (26/56) children with a postnatal hearing loss

- Formal factor analysis revealed a positive correlation between family history and postnatal hearing loss \( \chi^2(1) = 16.9, \ p<0.001 \)

- Logistic regression analysis revealed that children with family history as a risk factor were twice more likely to develop a postnatal hearing loss than those without family history (OR: 1.92; 95% CI: 1.04-3.56)
Syndromes

- Limited evidence
- Only one child with Branchio-oto-renal syndrome identified in the literature who passed the UNHS and identified with a postnatal hearing loss (Roth et al., 2008)
- This child also had preauricular skin tags and ear pits

Support for potentially monitoring
- Syndrome was present in 19.6% (11/56) of children with a postnatal hearing loss
- Formal factor analysis revealed a positive correlation between syndrome and postnatal hearing loss (\( \chi^2(1) = 32.2, p<0.001 \))
- Logistic regression could not be completed due to issues of multicollinearity with craniofacial anomalies
Prolonged Ventilation

- **Evidence not definitive** due to other complicating risk factors.
- Other risk factors include asphyxia, family history, and congenital diaphragmatic hernia (Masumoto et al., 2007, Robertson et al., 2009, Thiringer et al., 1984).
- For children who had received ECMO, 8.1%-12.6% of children developed a postnatal hearing loss (Mann et al., 1998, Fligor et al., 2005).

Support in favor of potentially monitoring:

- Prolonged ventilation was present in 19.6% (11/56) of children with a postnatal hearing loss.
- Factor analysis revealed a positive correlation between prolonged ventilation and postnatal hearing loss ($\chi^2(1) = 6.0, p=0.014$).
- Logistic regression could not be completed due to issues of multicollinearity with low birth weight.
Bacterial Meningitis

- **Limited evidence** as a substantial number of studies were excluded due to the exclusion criteria.
- Two studies included (Thiringer et al., 1984, Weichbold et al., 2006).
- Difficult to establish nature of the relationship given only children with a hearing loss were included and children had other risk factors.
- Limited information available.

Out of the 68 children referred for bacterial meningitis who passed UNHS, nil children developed a postnatal hearing loss.

**NB:** Other children in QLD have developed a hearing loss subsequent to bacterial meningitis during childhood. However, these children are excluded from analysis of a targeted surveillance program as they were identified due to medical referral.
Low Birth Weight

- Some evidence
- One high quality cohort study (Salamy et al., 1989)
- 6/224 children (2.7%) developed a postnatal hearing loss
- All 6 children had other contributing risk factors

Support in favour of not monitoring
- LBW present in 7.1% (4/56) of children with a postnatal hearing loss
- Formal factor analysis revealed a significant negative relationship between LBW and postnatal hearing loss
- Logistic regression analysis revealed that children with LBW as a risk factor were one-tenth more likely to develop a postnatal hearing loss than those with normal birth weight (OR: 0.14; 95% CI: 0.05-0.39)
Severe Asphyxia

- Some evidence
  - One high quality cohort study
  - 4/40 children (10%) developed postnatal hearing loss

Inconclusive evidence
- Severe asphyxia present in 16.1% (9/56) of children with a postnatal hearing loss
- Formal factor analysis revealed no significant correlation between severe asphyxia and postnatal hearing loss
Craniofacial Anomalies

- Limited evidence
- Weichbold et al., 2006
  - 2/23 children (8.7%) with a postnatal hearing loss had craniofacial anomalies
- Roth et al., 2008
  - 1/637 children (0.2%) with preauricular skin tags and ear pits developed a postnatal hearing loss. The authors recommended that children with skin tags and ear pits do not need to have their hearing monitored

Support in favour of monitoring
- Craniofacial anomalies present in 17.9% (10/56) of children with a postnatal hearing loss
- Formal factor analysis revealed a positive correlation between craniofacial anomalies and postnatal hearing loss ($\chi^{2}(1) = 5.4$, $p=0.020$)
- Logistic regression analysis revealed that children with craniofacial anomalies as a risk factor were more than two times more likely to develop a postnatal hearing loss than those without craniofacial anomalies (OR: 2.61; 95% CI: 1.19-5.70)
Limited evidence

One case report only was identified (Thiringer et al., 1984).

This child had other contributing factors including family history.

Hyperbilirubinemia was present in 3.6% (2/56) of children with a postnatal hearing loss.

Factor analysis revealed no significant relationship between hyperbilirubinemia and postnatal hearing loss.

POTENTIALLY DON’T MONITOR
Congenital Infection (TORCH)

- Evidence for CMV and toxoplasmosis only
- CMV
  - 1.3%-5.6% (asymptomatic) and 5.7%-14.4% (symptomatic) of children developed a postnatal hearing loss (Fowler et al., 1997, Iwasaki et al., 2007)
- Toxoplasmosis
  - No evidence associating toxoplasmosis to postnatal hearing loss (Brown et al., 2009)
- Limited evidence
  - Congenital infection present in 3.6% (2/56) of children with a postnatal hearing loss
  - Formal factor analysis revealed no significant relationship between congenital infection and postnatal hearing loss
Professional Concern

• Not explicitly reported in the literature
• May incorporate factors such as age, ototoxic therapy, and GA < 33 weeks
• Complex cases so difficult to know cause of the hearing loss
• No evidence
• No children (0/78) developed a postnatal hearing loss with professional concern
• Formal factor analysis revealed no significant correlation between professional concern and postnatal hearing loss

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