Success of Risk Indicators for Detecting Late Onset and Progressive Hearing Loss
An Analysis of the New Zealand Protocol

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Screening Programmes

“Screening programmes must continually re-evaluate protocols and procedures in order to maximise efficiency and use of resources and avoid burdening families with unnecessary appointments” Wood et al, 2013

Impact of targeted follow up protocol on audiology clinics in NZ is great due to the high percentage of referrals arising from the unique risk factors used
NZ data

Incidence of hearing loss in NZ

- Approximately 64,000 births,
- Universal new born screening roll out complete from 2010, international data suggests we should be finding 1-2/1000 or 60-120 infants/year
- NSU report for 6 months April 2011 – September 2011, 31,229 births, 18 identified or 0.56/1000 births
Increase in occurrence from birth to older childhood

• Numerous studies and data from screening programmes show an increase in hearing loss, presumably to progressive or late onset, however it is possible that some are mild losses that were not identified by screening.

• Due to the increasing duration of universal screening programmes in many countries it has now become possible to investigate the success of accepted risk factors in identifying late onset or progressive permanent hearing loss.


NZ Deafness Notification report shows pleasing increase in number of infants identified around the first few months from birth and then a second peak at age 4-5 due to the second universal hearing screen (B4 School check)

FIGURE 10: NUMBER OF CHILDREN DIAGNOSED BY AGE (2010 AND 2011)
Average age of identification for children with hearing loss at least moderate in degree over 15 years
Reporting Issues

• Deafness database report is dependent on Audiologist filling out the form and may be overlooked in the numerous forms that must be completed

• NSU report from 20 DHBs has acknowledged missing data in the audiology results. Also dependent on Audiologist filling out form

• Auditing of this feature of the screening programme not fully functional yet.

• (seem to have lost some babies with hearing loss somewhere, or else we are special in NZ!)
Identification of hearing loss

FIRST SUSPICION OF HEARING LOSS FOR CHILDREN BORN IN NZ (2012)
NZ risk factors for late onset and progressive losses

Largely based on JCIH 2007
• Family History
• Craniofacial anomalies
• Head trauma
• Bacterial/viral meningitis
• Syndrome
• TORCHS (suspicion not confirmation)
• Jaundice at the level of transfusion
• Ventilation (no duration specified)
• NICU more than 5 days (level 3)
• Other (often used for ototoxic drugs)
NZ largely adapted JCIH indicators with some minor modifications

Joint Committee on Infant Hearing (JCIH) 2007

Appendix 1. Risk Indicators Associated With Permanent Congenital, Delayed-Onset, or Progressive Hearing Loss in Childhood.

1. Caregiver concern regarding hearing, speech, language, or developmental delay (Roizen, 1999).
3. Neonatal intensive care of >5 days, or any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix), and hyperbilirubinemia requiring exchange transfusion (Fligor et al., 2005; Roizen, 2003).
4. In-utero infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis (Fligor et al., 2005; Fowler et al., 1992; Madden et al., 2005; Nance et al., 2006; Pass et al., 2006; Rivera et al., 2002).
5. Craniofacial anomalies, including those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies (Cone-Wesson et al., 2000).
6. Physical findings, such as white forelock, associated with a syndrome known to include a sensorineural or permanent conductive hearing loss (Cone-Wesson et al., 2000).
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome (Roizen, 2003). Other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson (Nance, 2003).
8. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome (Roizen, 2003).
9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis (Arditi et al., 1998; Bess, 1982; Biernath et al., 2006; Roizen, 2003).
10. Head trauma, especially basal skull/temporal bone fracture requiring hospitalization (Lew et al., 2004; Virtanen et al., 1985; Zimmerman et al., 1993).
11. Chemotherapy (Bertolini et al., 2004).
NZ protocol for targeted follow up

Pre October 2010
• Family History – Direct
• TORCHS for all
• Craniofacial, including pits and tags

Post October 2010
• Family History – immediate and second degree relatives – extended family members (blood relations), such as aunties, uncles, cousins and grandparents, included if known.
• Craniofacial Anomalies - exclusion of pits and tags in isolation
• CPAP - is no longer included in the NICU risk factors
• ECMO and IPPV remain as Ventilation Risk factors (no time limit)
• Risk factor questions asked of Well Babies - screeners are only required to ask the family history and phototherapy for jaundice questions. Screeners must continue to check for craniofacial anomalies including atresia, microtia and cleft palate (TORCHs dropped).
• Clarified ototoxic drugs had to be above therapeutic level
Protocol Changes
ADHB data April 2010 -
Targeted Follow Up Rates April 2011 - September 2011

20 DHBs
1 protocol, 20 interpretations
ADHB results

• ADHB screens approximately 14% NZ population due to large inter-DHB flows of high risk infants
• Provides audiology services for WDHB (NZ’s largest DHB) and ADHB, approximately 22% NZ population
ADHB All Risk Factors

Targeted Follow Up Risk Factors

- Cranio-facial
- Head Trauma
- Jaundice Requiring Transfusion
- Meningitis
- NICU level 3 >5 days
- Syndrome
- TORCHS
- Ventilation
- Other
- Family History
Family History
Due to very high numbers of referrals for TFU and the impact on audiology services the diagnostic test protocol was adapted to reduce the number of appointments required.

DPOAE screen at 18 months
If passing level of DPOAEs present in each ear – discharge
If not, continue testing with tympanometry and VRA

If strong concerns can see earlier
Success Of NZ Risk Factors
ADHB and WDHB

Data from April 2010 – October 2011
“Invitation to contact” – families are approached twice to ask for an appointment, if no contact made they are removed from the list
ADHB
- Notified Births 13,678,
- 643 referred for TFU
- Results for 275 (43% came for an appointment)
WDHB
- Notified Births approx 14,000
- Should have been 670 referred for TFU
- Results for 192 (difficult to calculate % as different database used)

No permanent hearing losses identified at 18 month point from approximately 28,000 births
Risk factors reviews from larger data sets


2,307,880 children born 01/04/06 – 30/09/09 in England. 2.99% of the birth population passed the screen with risk factors that required targeted surveillance.

The risk factors with the highest prevalence:

1. Syndrome (other than Down’s) associated with a hearing loss
2. NICU with refer in both ears at OAE and pass in both ears at AABR
3. Craniofacial anomaly
4. Down’s syndrome
5. Congenital infection
UK

- 98% offered out of 69043 eligible
- 53% took up offer of appt 38043
- 103 (0.35%) had a PCHI (30 unilateral and 73 bilateral)
UK

- Retain risk factors 1-5
- Others are discontinued
- “Not only is there a lack of robust evidence about the relative number of congenital PCHI and other later acquired PCHI in children, but additionally there is a lack of robust epidemiological data about the optimal age to look for later onset PCHI”.
- This strategy will be appropriate for programmes and countries where the prevalence and natural history of PCHI are similar to those in the UK.
Benchmark of <4% of children who are screened should be referred for targeted surveillance

Risk factors based on combination of JCIH and UK risk factors

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Note: If "Yes", Audiology surveillance is required; copy of completed form to be sent to Audiology.
Queensland Protocol

Pathway for Surveillance Refers:

- Infection
  - ABR at 3 months
  - VROA/BOA, OAEs and Tymps at 6 months
    - Normal VROA but unable to get separate ear info
      - Review every 6 months until 2 years of age
    - Abnormal
      - Diagnostic mode
      - WNL
      - Review every 6 months until 2 years of age
      - PTA at 3 years

- Family History
  - ENT recommendation regardless of hearing levels
  - Regular hearing review coordinated with ENT
    - Abnormal
      - Diagnostic mode
      - PTA at 3.5 years of age
      - Discharge from HH
    - Normal VROA but unable to get separate ear info
      - Review every 6 months until 2 years of age
      - PTA at 3 years

- Down Syndrome Or Cleft Palate
  - VROA/BOA, OAEs and Tymps at 9-12 months
    - Other 8 risk factors
      - Normal VROA but unable to get separate ear info
        - WNL
      - Discharge from HH

[where BOA = Behavioral Observation Audiometry; ENT = Ear, Nose, Throat specialist; HH = Healthy Hearing Program; info = information; OAEs = Otoacoustic Emissions; Tymps = tympanometry; PTA = Puretone Audiometry; VROA = Visual Reinforcement Observation Audiometry; WNL = Within Normal Limits]
Queensland

- 7320 children for targeted surveillance (2.8% of 261,328)
- 97.3% had normal hearing who completed the very intensive appointment series and discharged from the program
- 56 (0.77%) had hearing loss
- Yields for risk factors (from total 7320)
  - Syndrome 3.1%
  - Craniofacial 1.7%
  - Severe asphyxia 1.5%
  - Family history 0.9%
  - Prolonged ventilation 0.6%
Common findings

degree of hearing loss detected:

• half of the cohort was identified with a mild degree of hearing loss.

• “As mild hearing losses may not be detected at the newborn hearing screen, it is possible that the hearing loss was present at birth in these children”.
Problems with ongoing surveillance

- Poor attendance
- Cost to families of both time and money to attend appointments
- Burden on audiology and administration services
- Reduction in adult services to accommodate requirements of screening programme
Impact on audiology if UK protocol adapted

Numbers would drop to approximately 5 per month
NZ pathway?

- Universal Newborn Screen
- Universal B4 School Screen
- Targeted surveillance of children with risk factors by B4 School check (would require national database to be effective)
- Door always open to referrals from GP and other health professionals over hearing concerns
- Many years before national data available
- Follow UK guidelines?