

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

Wood, Davis, Sutton, 2013. *International Journal of Audiology*, 52: 394–399

Beswick, Driscoll, Kei, Khan, Glennon, 2013. *J Am Acad Audiol* 24:205–213

Beswick, Driscoll, Kei, Glennon, 2012. *International Journal of Pediatric Otorhinolaryngology* 76 , 1046–1056

Deafness Notification Report (Digby, 2013)

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)

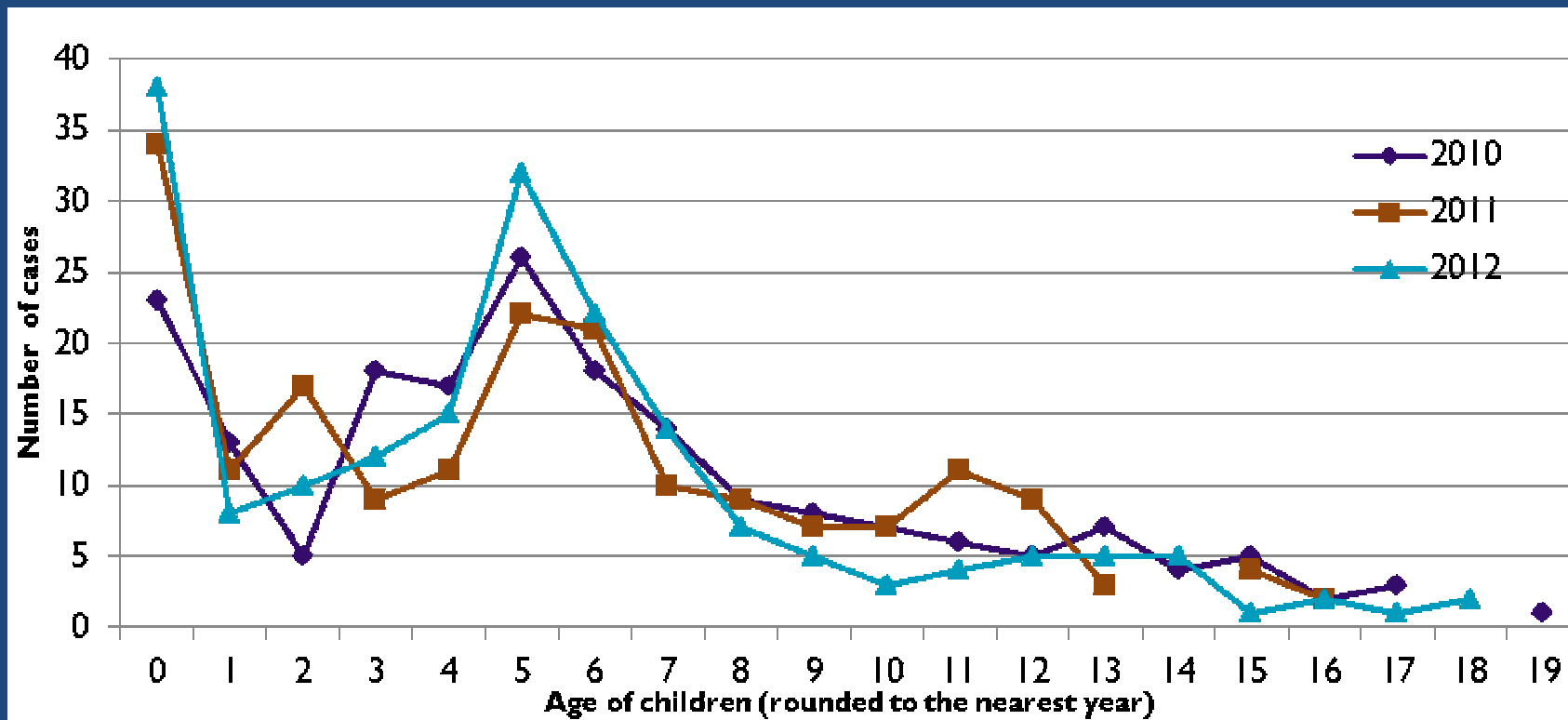
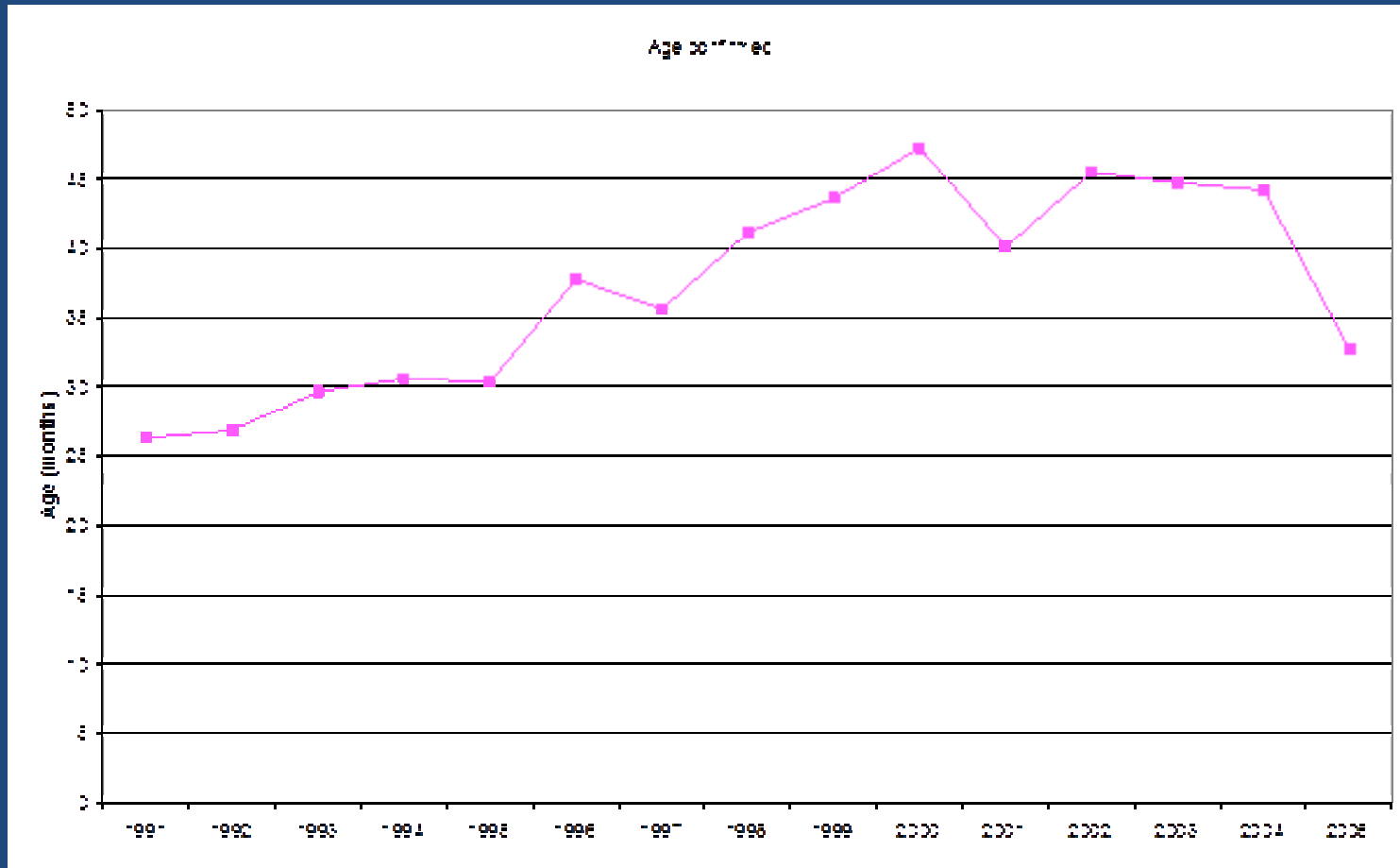


FIGURE 10: NUMBER OF CHILDREN DIAGNOSED BY AGE (2010 AND 2011)

Deafness database 2005

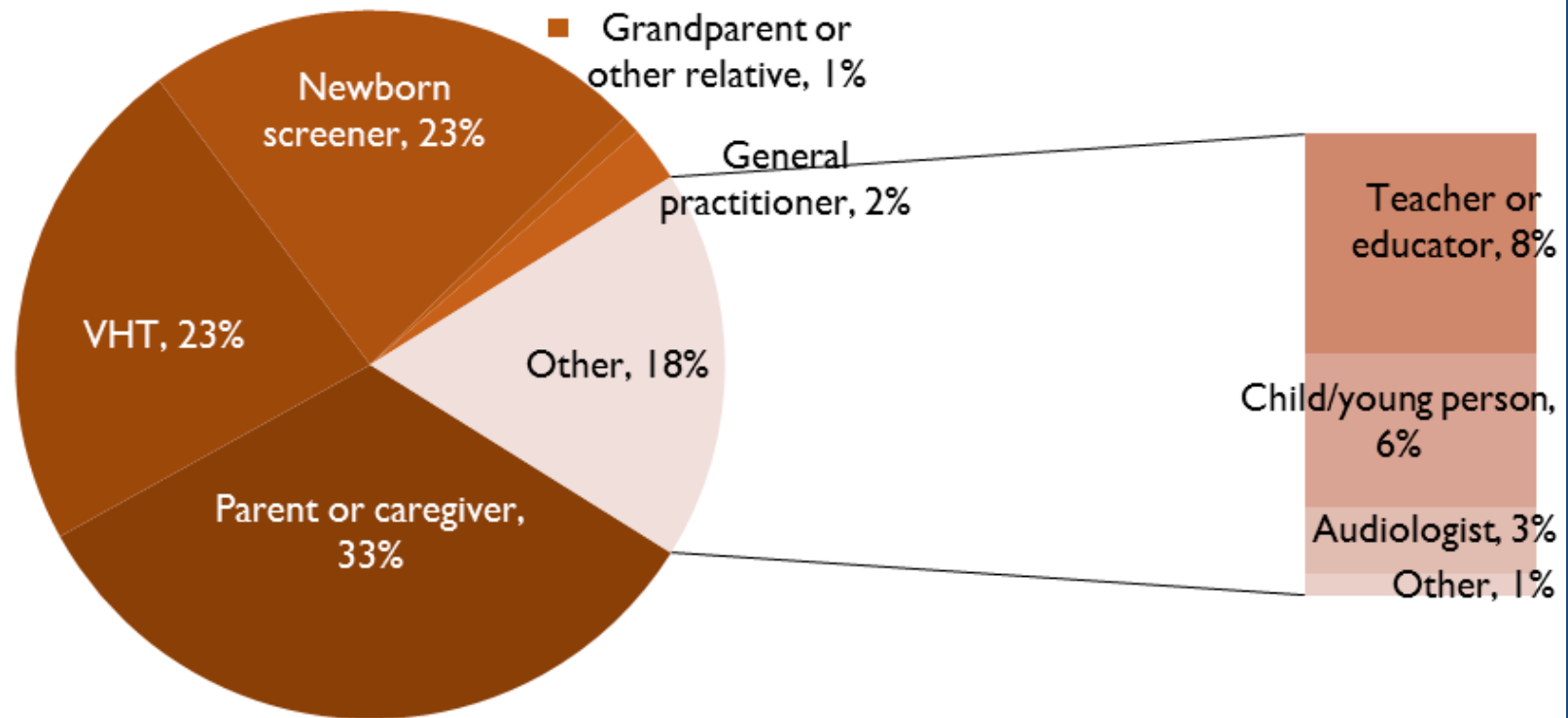


Average age of identification for children with hearing loss at least moderate in degree over 15 years

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^a regarding hearing, speech, language, or developmental delay (Roizen, 1999).
2. Family history^a of permanent childhood hearing loss (Cone-Wesson et al., 2000; Morton & Nance, 2006).
- 3. Neonatal intensive care of >5 days, or any of the following regardless of length of stay: ECMO,^a 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,^a 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,^a 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,^a such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome (Roizen, 2003).
9. Culture-positive postnatal infectious associated with sensorineural hearing loss,^a 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^a requiring hospitalization (Lew et al., 2004; Vartiainen et al., 1985; Zimmerman et al., 1993).
11. Chemotherapy^a (Bertolini et al., 2004).

NZ protocol for targeted follow up

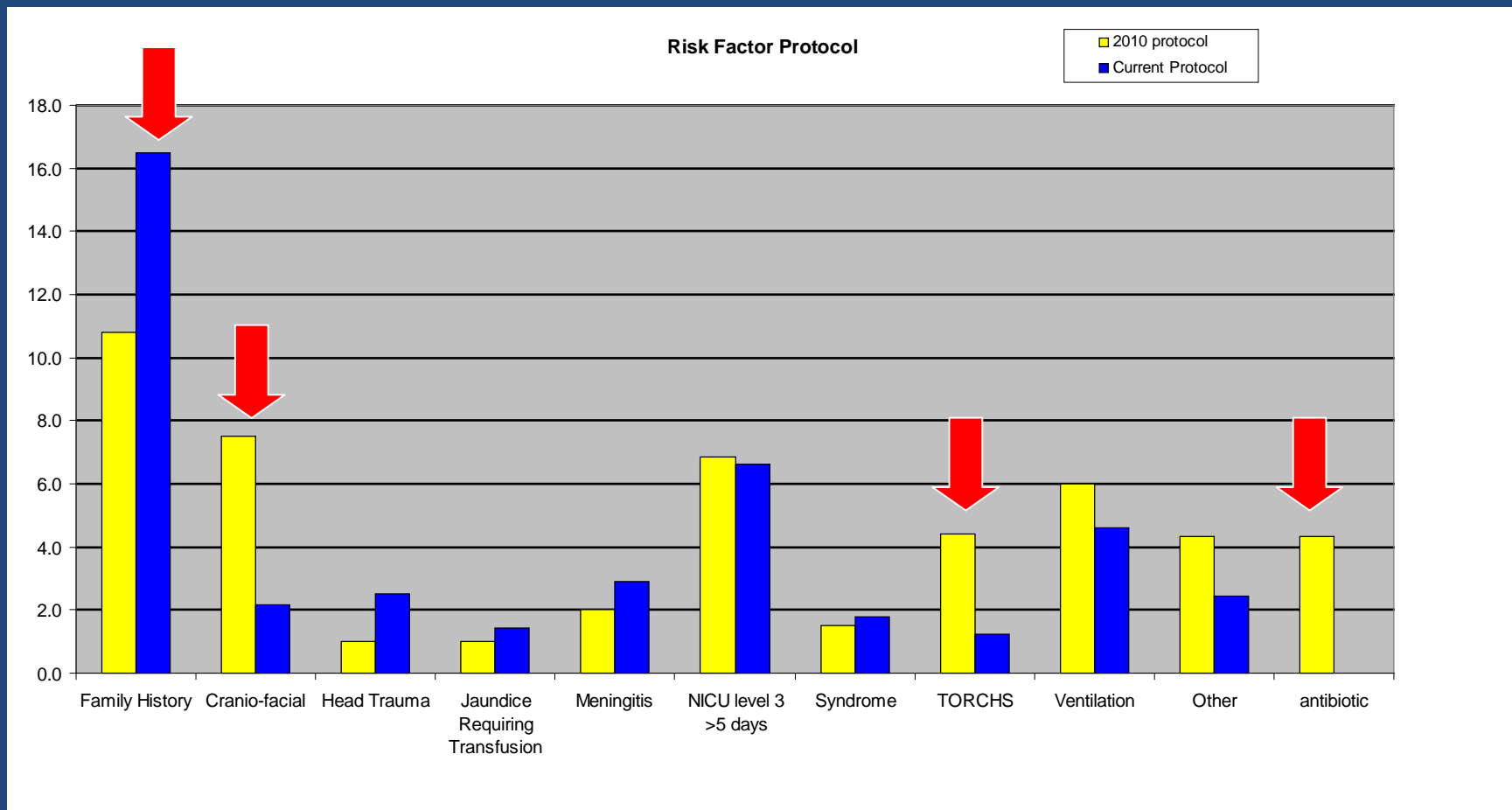
Pre October 2010

- Family History – Direct
- TORCHS for all
- Craniofacial, including pits and tags

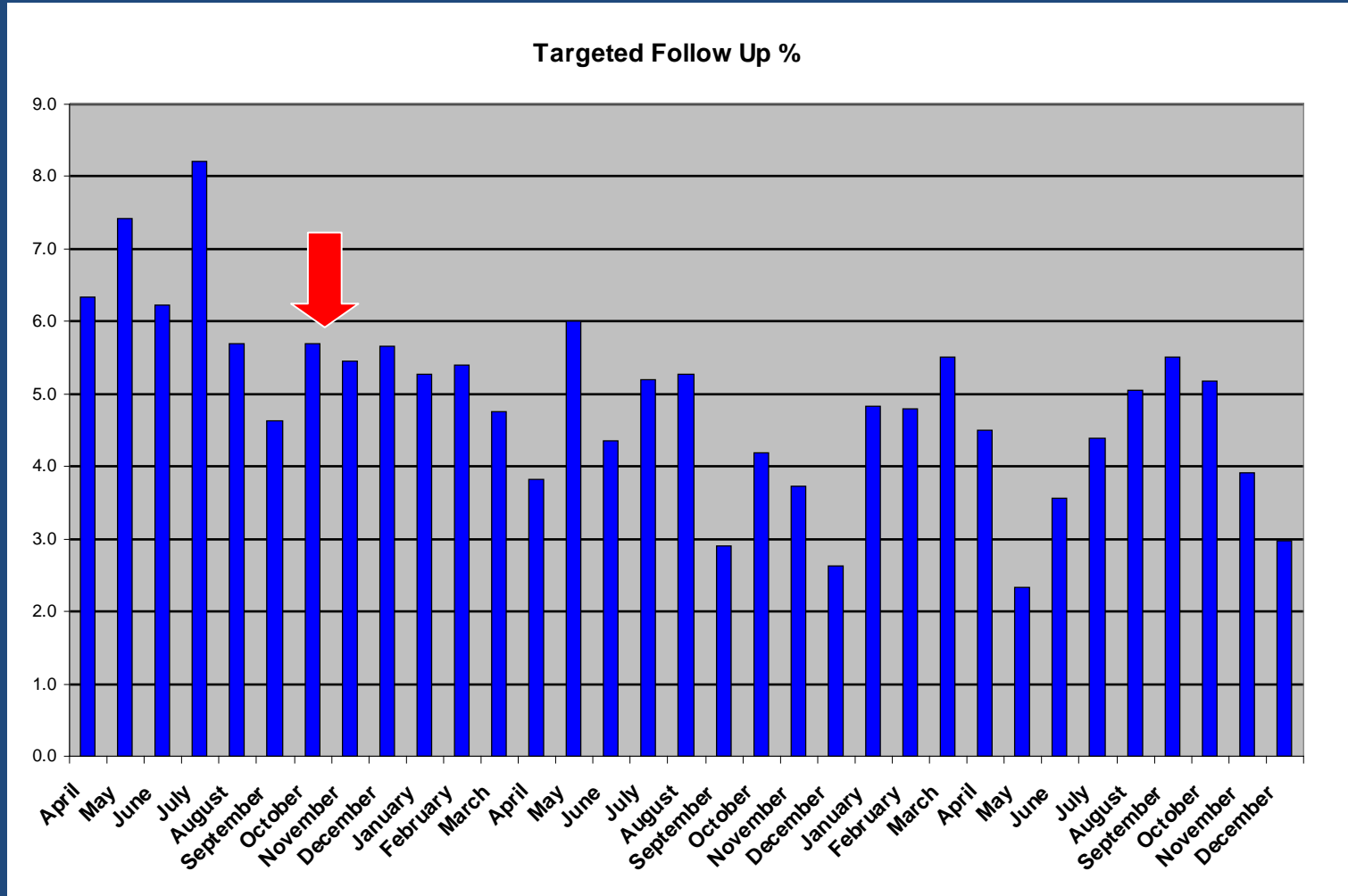
Post October 2010

- Family History –immediate and second degree relatives –extended family members (blood relations), such as aunts, 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

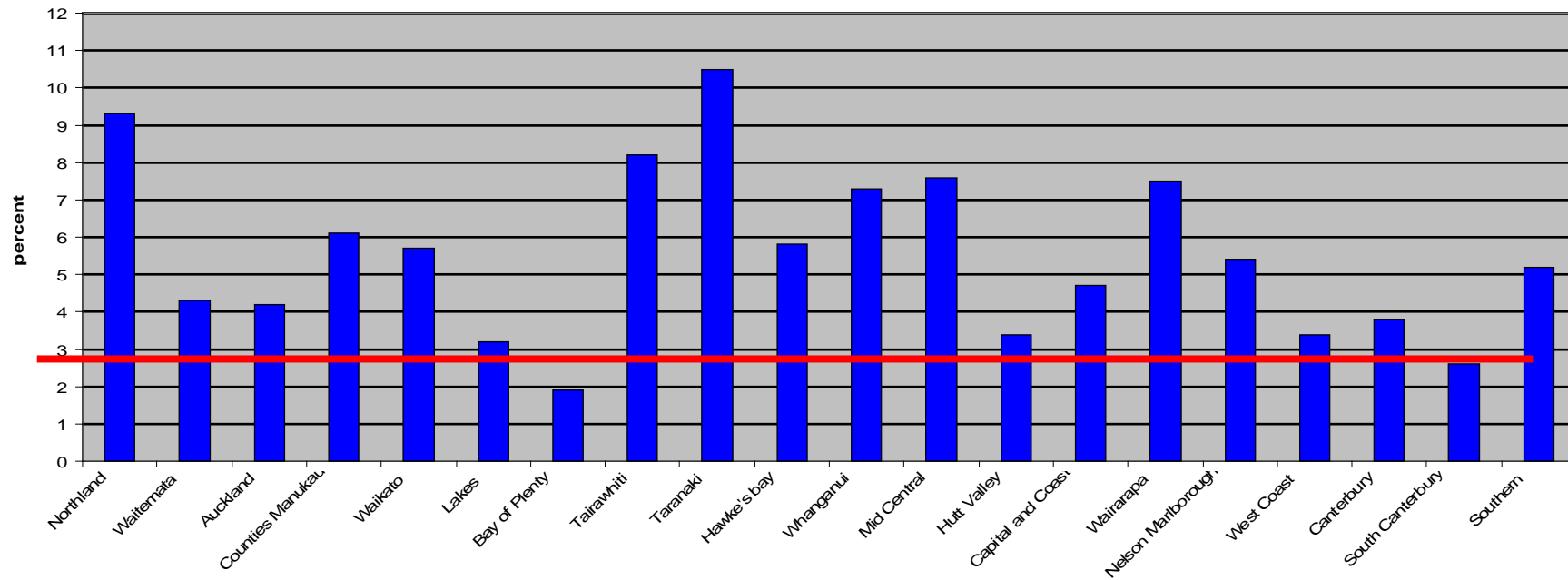
Protocol Changes ADHB data April 2010 -



ADHB



Targeted Follow Up Rates April 2011 - September 2011



20 DHBs

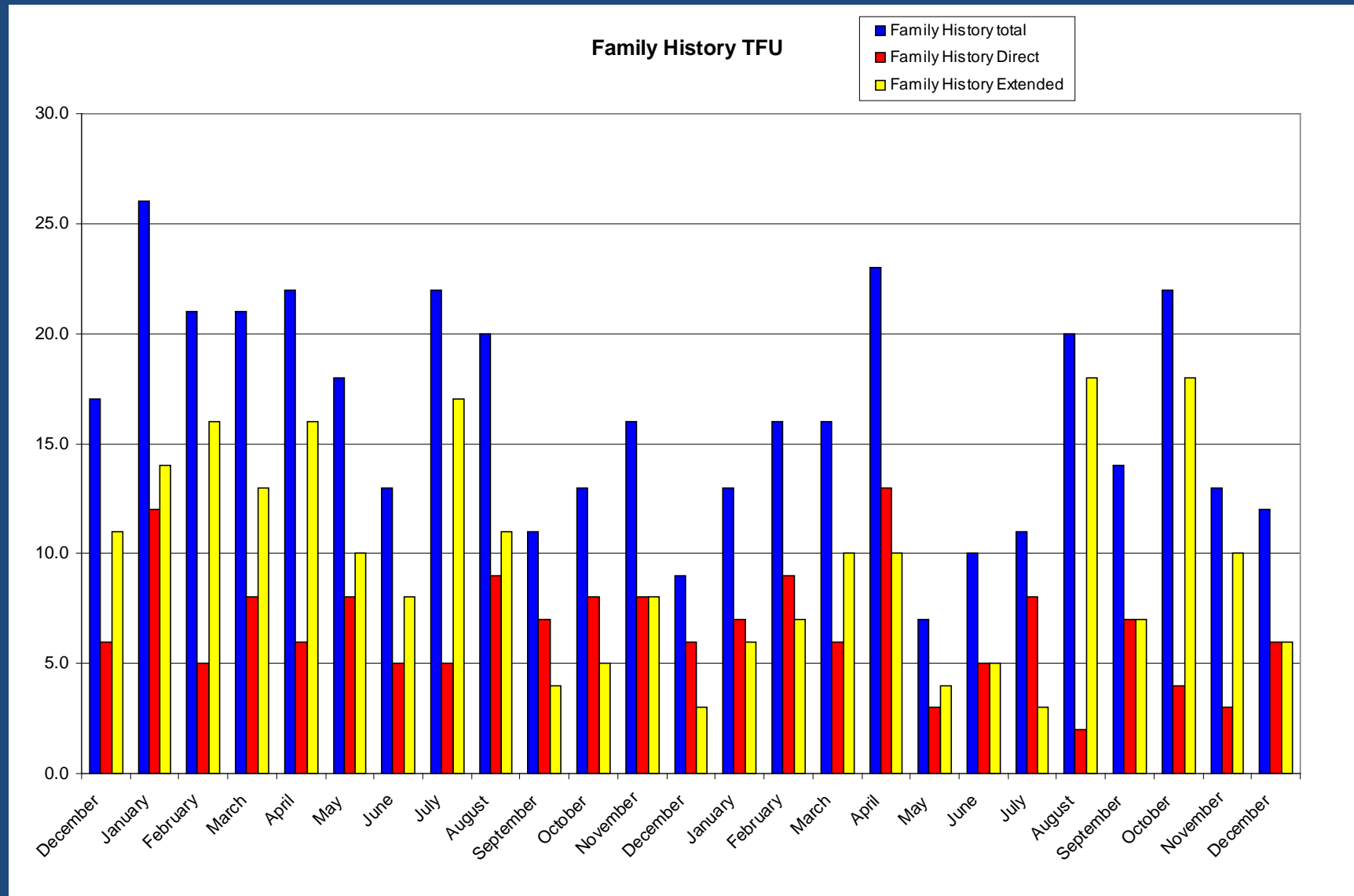
1 protocol, 20 interpretations



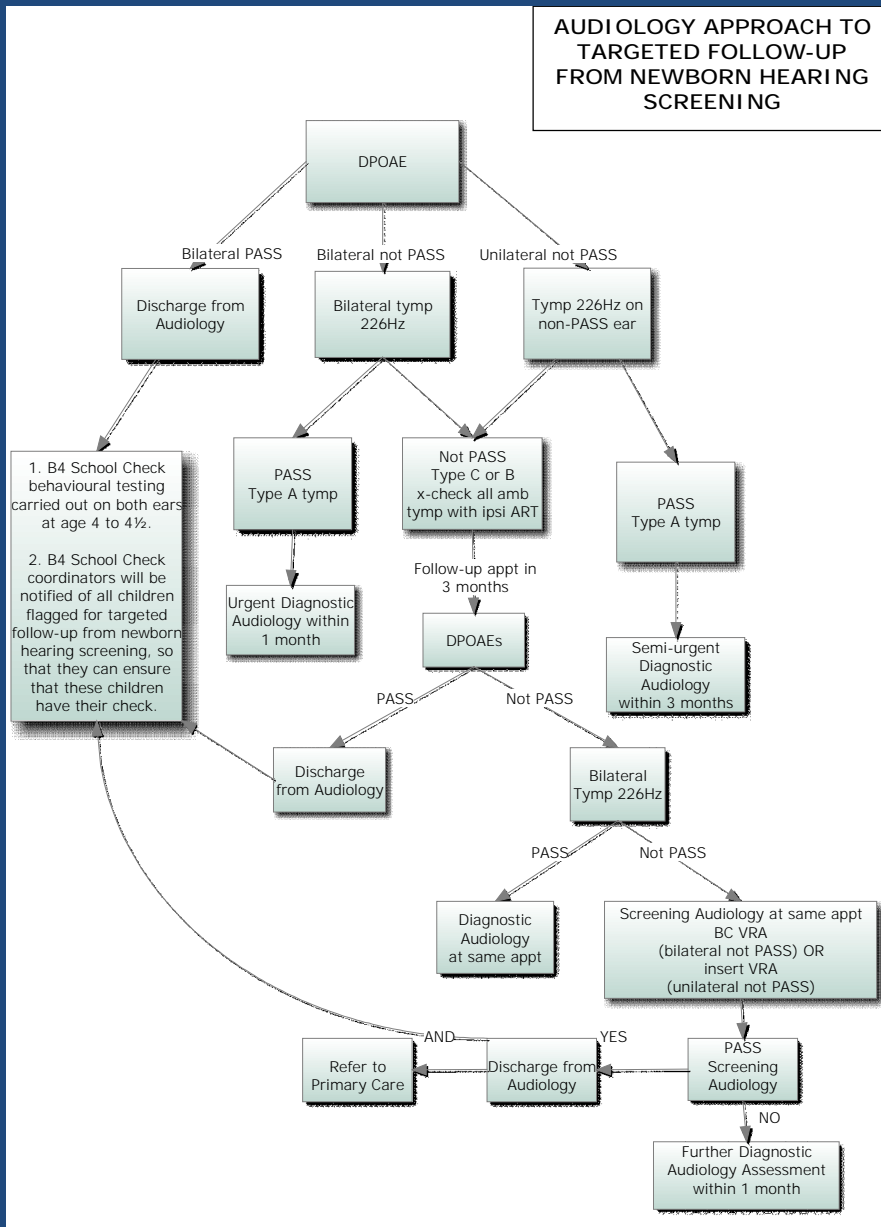
ADHB results

- 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

Family History



Current Audiology Targeted Follow Up Protocol



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

UK Results (Wood, Davis, Sutton, 2013.

International Journal of Audiology, 52: 394–399)

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.

Queensland

Beswick, Driscoll, Kei, Glennon, 2012. International Journal of Pediatric Otorhinolaryngology 76 , 1046–1056

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

High Risk Indicators (If 'Yes', Audiology surveillance is required: copy of completed form to be sent to Audiology)

Yes No Family history of permanent childhood hearing loss (mother / father / siblings of baby only) excluding grommets / ear infection / trauma
Details:

Yes No Syndromes associated with hearing loss (eg. Downs, FAS) Details:

Yes No Prolonged ventilation \geq 120 hours (IPPV / CPAP / HHFNCT) Number of hours:

Yes No Bacterial meningitis (confirmed / suspected) Details:

Yes No Low birth weight \leq 1500 grams Weight: g

Yes No Severe asphyxia at birth (convulsions / HIE / PPHN) Details:

Yes No Craniofacial anomalies eg. cleft palates (excluding cleft lips and skin tags) Details:

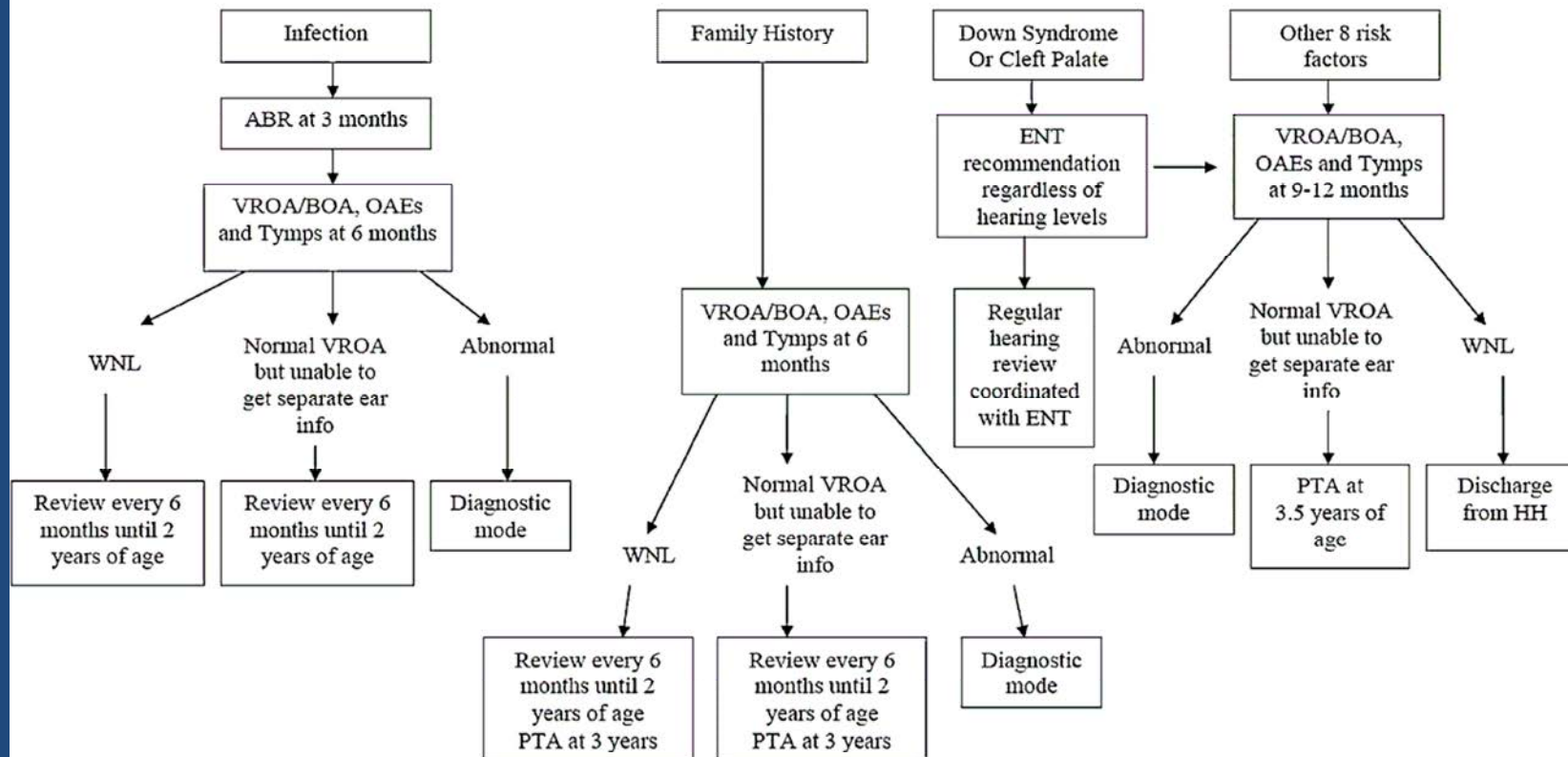
Yes No Hyperbilirubinemia levels \geq 450 μ mol/l (Term) or \geq 340 μ mol/l (Preterm) Max SBR level: μ mol/l

Yes No Proven / suspected congenital infection of the baby Toxoplasmosis Rubella CMV Herpes Syphilis

Yes No Professional concern Details:

Queensland Protocol

PATHWAY FOR SURVEILLANCE REFERS:



[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

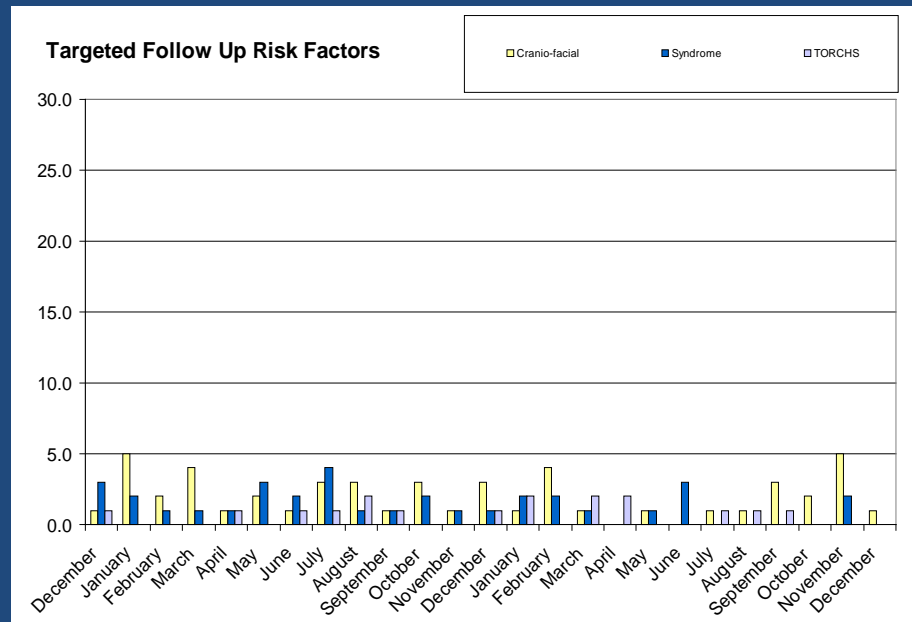
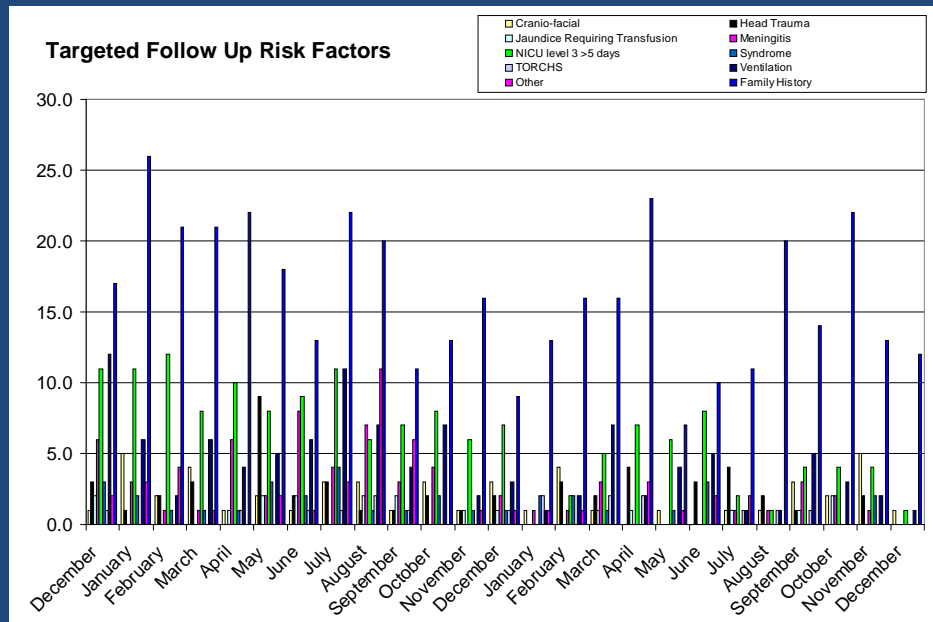
degree of hearing loss detected:

- 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 on going 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?