

# When a Unilateral refer reveals a Bilateral loss: cause for concern?

Auckland May 2013

Children's Hospital at  
Westmead



Photo courtesy of Anne Porter,ADK

## Global program attitudes to unilateral loss

- Chang (J.MedScreen.2009) reviewed 66 papers on NHS 2000-7 :
  - \*15 articles considered unilateral refers to be passes
  - \*17 combined unilateral and bilateral as one refer rate,
  - \*34 separated unilateral and bilateral and demonstrated follow-up for separate ears
- 'Differential treatment of unilaterals persists today in some areas of the world'
- Her study - 'up to 40% of infants(without aural congenital abnormalities) who pass in one ear and fail in the other may actually have bilateral loss'
- How does this happen?

# SWISH - AABR screening trade offs

Intensity \*

- ALGO Template is patterned on responses of infants with thresholds of 0-15dB (i.e. softer than 35db presentation level)
- AABR is a *supra-threshold* response.
- An infant with a loss of 25dB will refer, because this will alter the height and length of wave peaks : response won't match the template.

\* Taken from Natus Clinical Series No.3

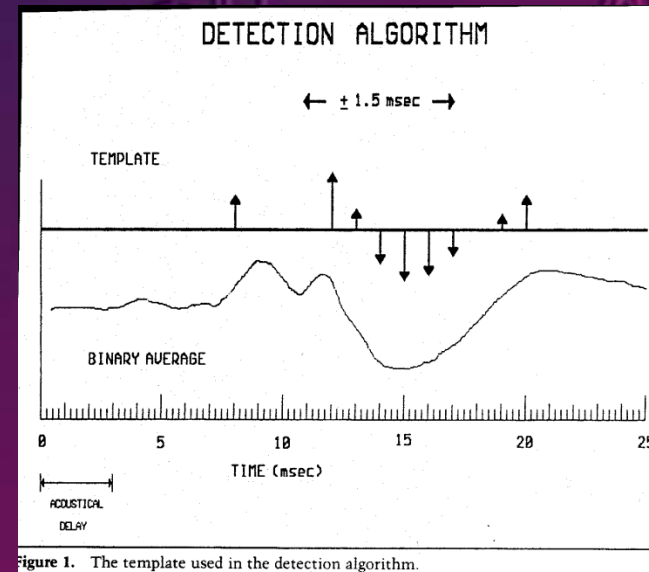
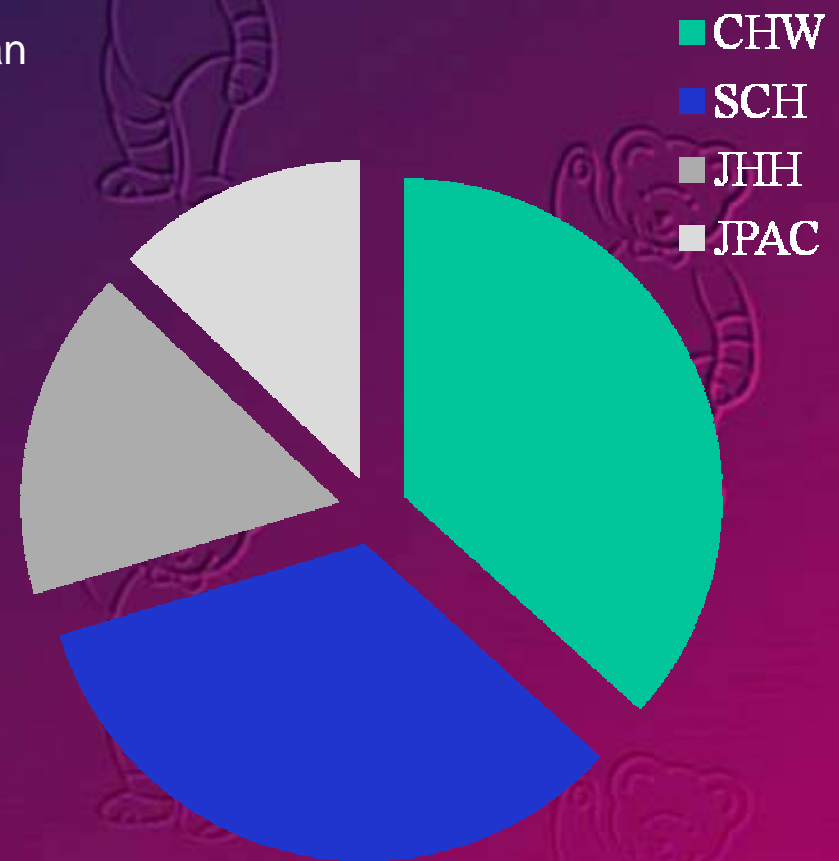


Figure 1. The template used in the detection algorithm.

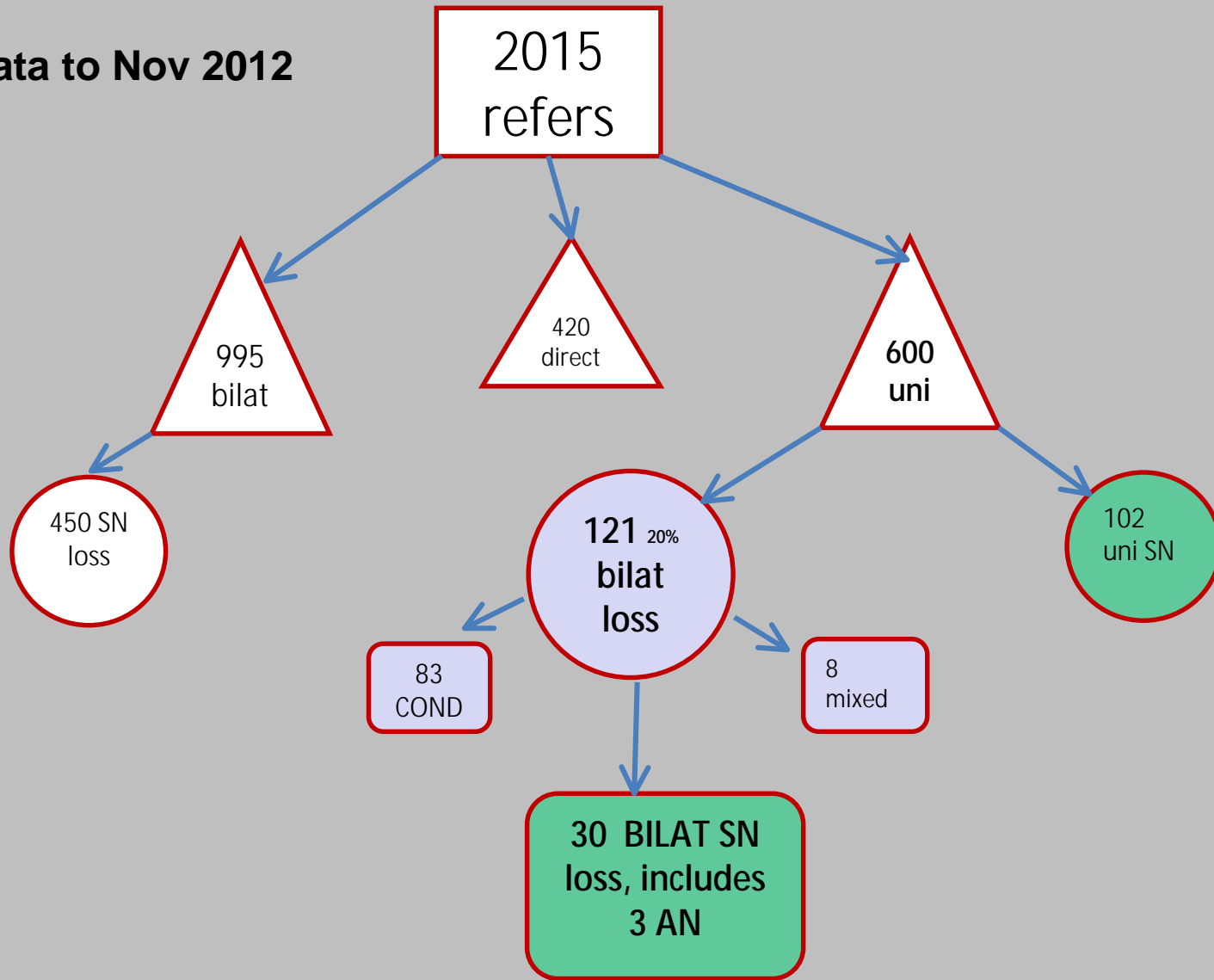
Frequency

Broadband signal, like a piano chord(700-5Khz).One or some of the frequencies *might be missing*, but this can be masked by strength of peak signal in a different part of the spectrum, producing a pass.

- SWISH Program began late 2002, now screens 98,000 births per year, over 1/3 of total Australian births.
- Diagnostic referrals shared by 4 clinics (Referral rate ~0.8%).
- CHW break-up for 2010-11 was 108 bilateral refers, 101 unilateral, 88 'direct'.
- CHW unilaterals BIASED towards sicker, higher needs, high risk infants (access to paed/sw/ent/imaging etc)
- JPAC takes low risk (in theory) Western Sydney unilateral overflow of another 104 unilaterals.



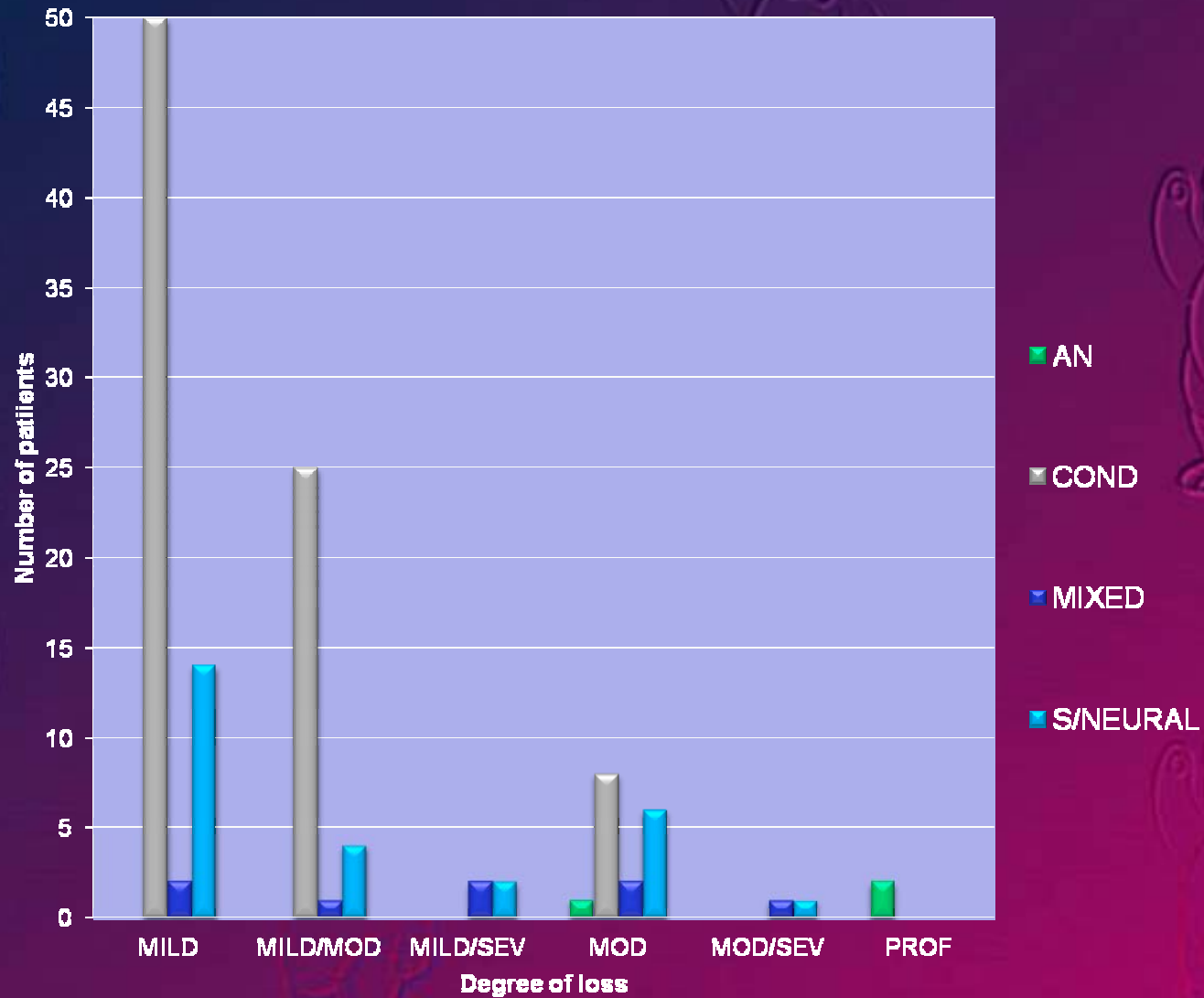
**CHW Data to Nov 2012**



## Bilateral loss from unilateral refer



## Type and level of loss



<b>Risk factor for 83 conductive</b>	<b>Number (not exclusive)</b>
<b>Craniofacial(mainly cleft)</b>	<b>33</b>
<b>NIL</b>	<b>23</b>
<b>NICU</b>	<b>12</b>
<b>Down Syndrome</b>	<b>9</b>
<b>Prematurity</b>	<b>8</b>
<b>Family history</b>	<b>4</b>
<b>Achondroplasia</b>	<b>1</b>
<b>Spina Bifida</b>	<b>1</b>
<b>Cardiac anomalies</b>	<b>1</b>
<b>Bilat.stenotic canals</b>	<b>1</b>

## **CONDUCTIVE LOSS**

- Comprise 70% of unilateral refer to bilateral losses
- Almost all transient, monitored via Cleft Palate Clinic, including 7/8 of the moderate losses
- Only 1 moderate loss fitted-stenotic canals



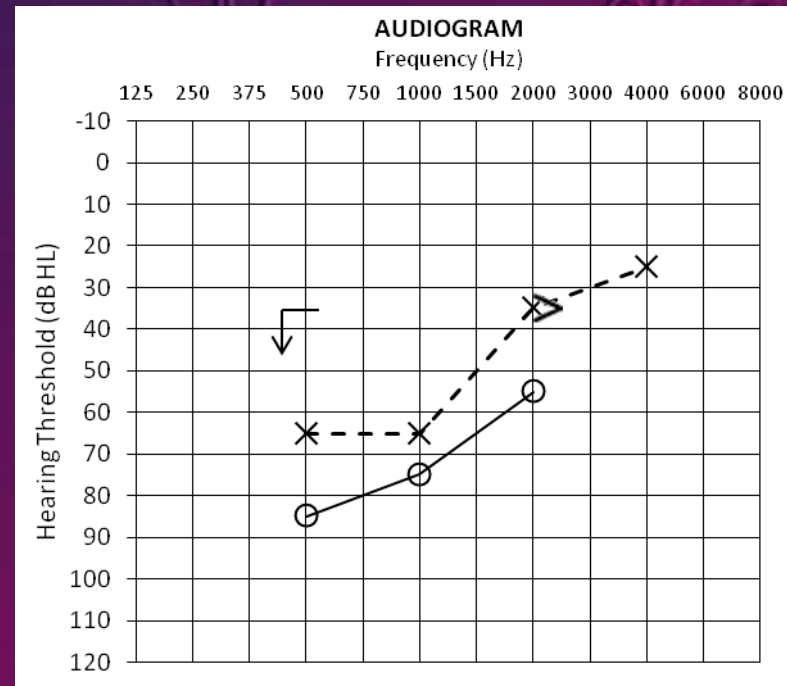
<b>Risk factors for 8 mixed loss</b>	<b>Number (not exclusive)</b>
<b>Nil</b>	<b>1</b>
<b>NICU placement</b>	<b>3</b>
<b>CFA</b>	<b>2</b>
<b>CMV</b>	<b>1</b>
<b>Family Thalassemia</b>	<b>1</b>
<b>Hypothyroid</b>	<b>1</b>
<b>Premature</b>	<b>1</b>
<b>Jaundice</b>	<b>2</b>
<b>Talipes(foot deform.)</b>	<b>1</b>

## **MIXED LOSS**

- 3 were fitted binaurally with hearing aids
- 5 were monitored with tympanometry, OAE and later VROA

# Infant 1: mixed

- Pass left ear/refer right ear
- Type A's both ears
- Absent OAE's
- Multiple congenital abnormality
- Fitted bilaterally with hearing aids



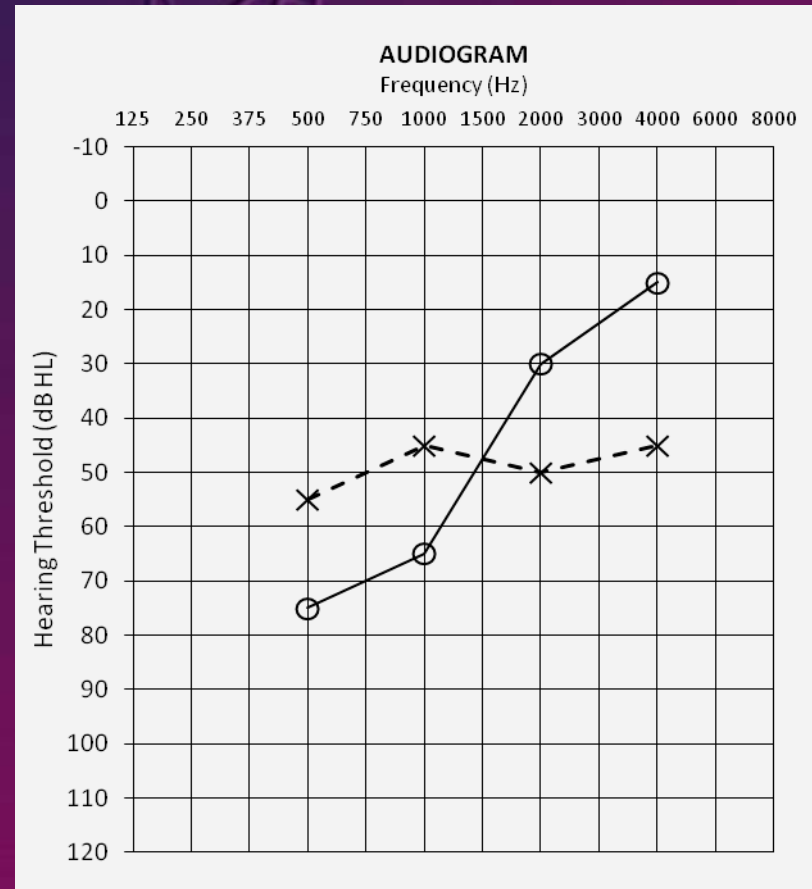
Risk factors for 27 <i>sensori-neurals</i>	Number (not exclusive)
Nil	8
NICU placement	11
Connexin	2
Dysmorphic	2
Family History	2
Large Vestibular Aqueduct	2
Premature	1
CHARGE	1
Pierre Robin	1
Twin	1
Dysplasia	1
Hypothyroid	1
Jaundice	1
Stickler	1

## SENSORINEURAL LOSS

- Comprises 6% of total binaural SN loss detected, 5% of total unilateral refers
- 19 fitted bilaterally with hearing aids (70%)
- 7 not – very mild/borderline
- 1 lost to follow-up at Australian Hearing

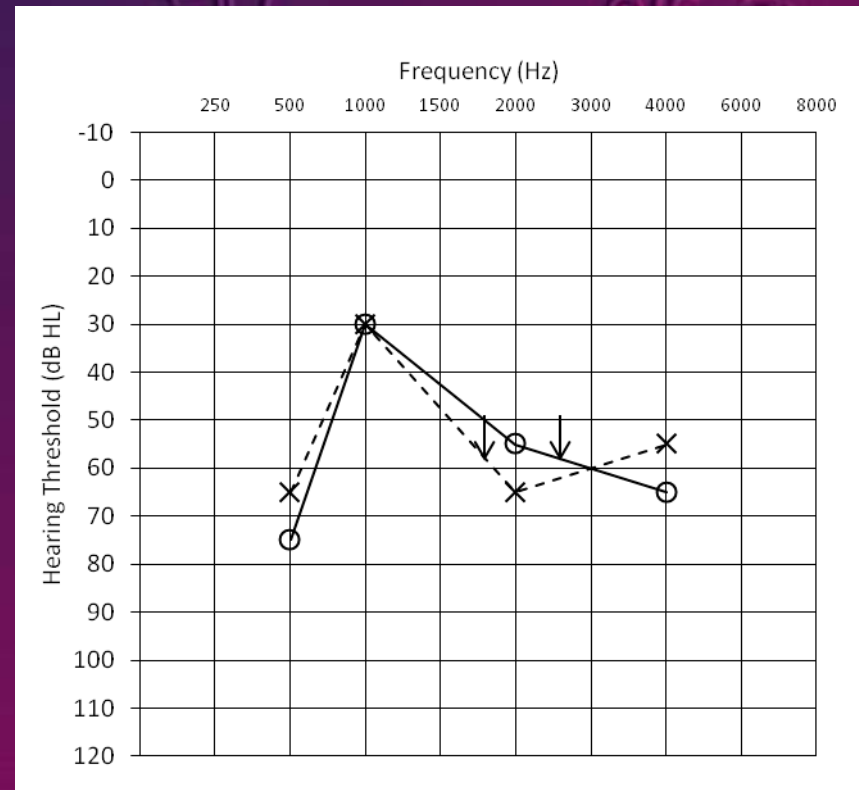
## Infant 2 :Sensorineural

- Pass right ear/refer left ear
- Type A's both ears
- Nogo bone conduction  
Emissions absent both ears
- Risk factors ++  
prem/VSD/Dysmorphic
- Fitted bilaterally with hearing aids

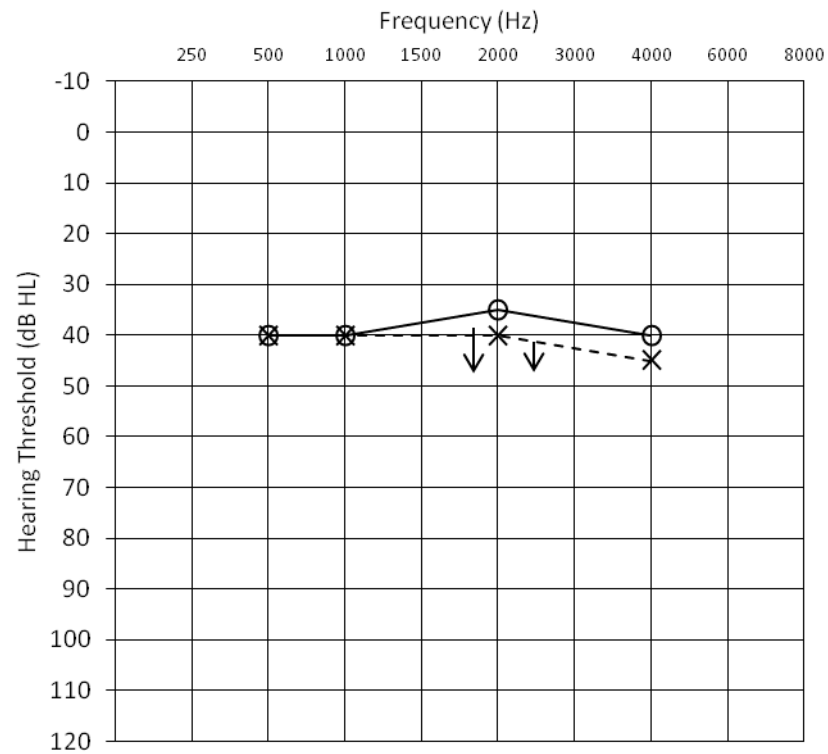


# Infant 3 :Sensorineural

- Pass right ear /refer left ear
- Type A's both ears
- Emissions absent both ears
- No known cause - query respiratory distress but not confirmed
- Fitted bilaterally with hearing aids



## Case Study : Infant 4



- Pass right ear /refer left ear
- Normal tymps
- Absent emissions
- No risk factors, sister has syndrome but no hearing loss
- Fitted bilaterally with hearing aids

# Psycho-social impact of detection of bilateral loss

## First child

- 2 year old sister also patient at CHW
- Heart surgery soon after birth –NICU/PICU, ongoing learning and cleft issues
- We diagnosed bilateral conductive at birth ABR.Tubes during cleft surgery
- So not concerned- imagined this referral also due to transient unilateral conductive
- Felt 'safe' coming to CHW

## This child

- Bilateral permanent loss signalled loss of idealised second child and arrival of new set of issues
- 'already gone through a lot'
- Intense initial phone counselling by our Social Worker, then regular calls for 6 months.
- GP also arranged local counselling.

# How do bilateral AN's get through?

Patient	Ear	AABR	T	O	0.5	1K	2K	4K	CM Rev	HA CI	Risk factors
1	R	refer	A	Abs	-	nr85	nr85	nr85	yes	yes	28/40, NICU, PDA
	L	PASS	A	Abs	-	nr85	nr85	nr85	yes	yes	

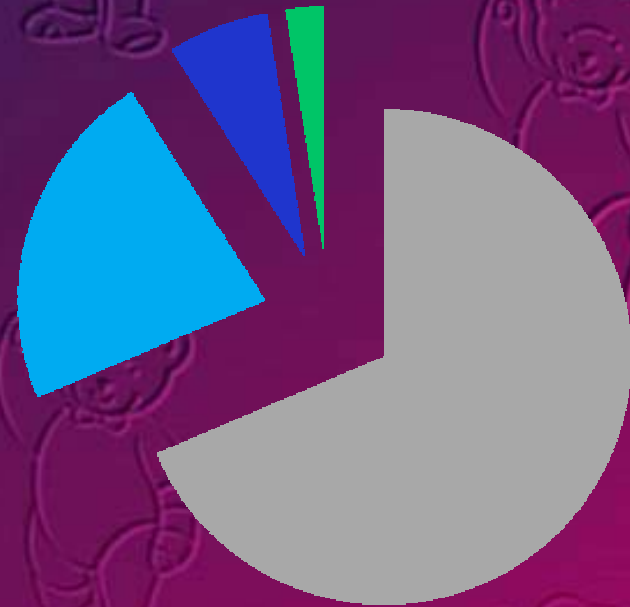
2	R	refer	A	Pres	-	nr85	nr85	nr85	yes	yes	Jaundice, hypothyroid
	L	PASS	A	Pres	Click 15	35	45	55	yes	yes	

3	R	PASS	A	Abs	60	45	55	65	yes	yes	29/40, twin PDA, Chronic lung disease
	L	refer	A	Abs	60	45	55	65	yes	yes	



# Conclusions

- Not alarming, but surprising
- Large percentage of risk factors unexpected
- Needs to be an awareness both at screening and diagnostic level that 1:5 of our unilateral refers will reveal a bilateral loss
- 1:20 will be bilateral PHL
- Recognition of impact on family, even if conductive : results vs expectation
- Require similar levels of psychosocial and medical after care/ investigation as bilaterals, including genetics/parent to parent



Question : could an OAE/AABR protocol\* be modified to improve identification of these cases?

\*See Jean Johnson 2005 : 23% of PHL kids at 9m had failed OAE but passed AABR.

### *Limitations of Study*

- Our sample of unilaterals is biased
- Only 40% of SWISH sample
- SWISH not deemed worthy enough to deserve a database, so we use a homegrown Access database: errors, missing data, inconsistent entries.

### CHW Audiology team



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