

# Prevalence and parental awareness of hearing loss in children with Down syndrome

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## • Down syndrome

- 1 in 700 live births in Hong Kong
- Hearing loss is found in 38 to 78% of patients with DS
- Data on hearing impairment of DS in Chinese population is scarce
- Hearing evaluation in DS is difficult
  - inconsistent voluntary response and poor attention during behavioral testing
- **BAEP**: high sensitivity and specificity (>90%) and can evaluate auditory performance regardless of patient's age and mental status
  - Advocated by **AAP** (2011) to establish hearing status in DS

## Objectives:

### Primary :

- **To identify the point prevalence and types of hearing loss in a cohort of ethnic Chinese DS children** actively followed up in a specialized clinic for DS patients at a regional paediatric centre.

### Secondary :

- To determine the **degree of agreement between BAEP findings and parental awareness of hearing loss**

# Method

- A **cross-sectional** study
- **Subjects:** DS children followed up in the Down Syndrome Clinic at Caritas Medical Centre with age ranged from 0 to 18 years
- Diagnosis of DS confirmed by karyotype studies
- A **structured questionnaire** was used to obtain participant's case history
  - Symptoms of hearing impairment were classified into 4 groups (normal, mild, moderate, severe)
- Concomitant **BAEP study** performed within 12 months was retrieved for analysis
- **Data analysis** : SPSS version 12.0 , MedCalc Statistical Software version 12.7.2 for calculating weighted kappa

<b>Gender</b>	
Male	35 (70%)
Female	15 (30%)
<b>Age (years)</b>	
Mean	11.7 (SD +/-5.7)
Median	13.9
<b>Intellectual disability</b>	
Mild	23 (46%)
Moderate	22 (44%)
Unavailable	5 (10%)
<b>History of Otitis media</b>	
Yes	6 (12%)
No	44 (88%)
<b>Use of hearing aids</b>	
Yes	1 (2%)
No	49 (98%)
<b>Active Follow up in ENT</b>	
Yes	13 (26%)
No	37 (74%)

# Results

- The **point prevalence** of hearing loss is estimated to be **36%**, which is similar to a recent population-based study in Norway (35%)
- Hearing threshold ranged from 30 to 70 dBnHL
  - **Conductive deficit** : 13 patients (**72.2%**)
  - **Sensorineural deficit**: 5 patients (**27.8%**)

BEAR results		Number
Normal		32 (64%)
<b>Conductive deficit</b>		<b>13 (26%)</b>
	Mild (20-40dBnHL)	6 (46.1%)
	Moderate (≥40-60dBnHL)	2 (15.4%)
	Severe (> 60dBnHL)	5 (38.5%)
<b>Sensorineural deficit</b>		<b>5 (10%)</b>
	Mild (20-40dBnHL)	1 (20%)
	Moderate (≥40-60dBnHL)	2 (40%)
	Severe (> 60dBnHL)	2 (40%)
Total subjects with hearing loss		<b>18 (36%)</b>
	Bilateral	8 (44.4%)
	Unilateral	Left 8 (44.4%) Right 2 (11.2%)

## Parents perception:

- Care takers of 13 patients (26%) perceived their children had symptoms of hearing impairment
  - Mild: 9 (69.2%)
  - Moderate: 3 (23.1%)
  - Severe: 1 (7.7%)

Inter-rater agreement between BAEP findings and symptoms of hearing impairment:

- **Quadratic weighted kappa = 0.045**  
(95%CI -0.138 to 0.229)

→ **very poor strength of agreement** between BAEP and clinical questioning in identifying the degree of hearing loss

Degree of hearing problem by questionnaire	Severity of Hearing deficit estimated by BAEP				Total
	Normal	Mild	Moderate	Severe	
Normal	25	5	<b>3</b>	<b>4</b>	37
Mild	4	1	<b>1</b>	<b>3</b>	9
Moderate	3	0	0	0	3
Severe	0	1	0	0	1
Total	32	7	4	7	50

**Quadratic weighted Kappa = 0.045(95% CI -0.138 to 0.229)**

# Conclusion

- Estimated point prevalence of hearing impairment in Chinese DS children in Hong Kong is 36%
- Poor agreement between BAEP findings and parental reporting of hearing problems reflects poor parental awareness of hearing problems among DS patients
- Early detection of hearing problem is crucial to language development and continuous surveillance of hearing is mandatory for DS patients
- BAEP is a useful objective test in evaluating hearing status regardless of patient's age and mental status; and is helpful in uncooperative children who are not suitable for behavioral audiogram or PTA. (e.g DS patients)