

Cochlear Implant in children with additional disabilities

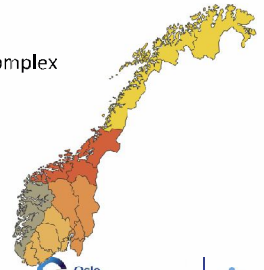


Marie Bunne
MD, PhD, Surgeon, Section for Otology
Oslo University Hospital



Rikshospitalet

- The only centre for children in Norway, 4.5 million people
- CI in children < 6 years since 1988
- Bilateral CI in children since 2004
- CI in prelingually deaf children with complex handicaps since 2005
- ~ 25-30% with additional handicap



Population

- 70 out of totally 397 implanted children 1995 – 2008 in Norway (~18%)
 - Age at first CI:
 - mean 3 yrs 9 months (5 m -1.5 yrs), median 2 yrs 9 m
 - 48 (69%) bilateral
 - 43 Cochlear (Nucleus), 27 MEDEL
- Follow-up time:
 - mean 6 yrs 9 months (10 m – 17 yrs), median 6 yrs 3 m



Difficulties in definition & categorisation

- Clinical impairment/disability:
 - Varying etiology
 - May / may not have cognitive disabilities
- Syndromes:
 - Varying severity
 - May / may not have cognitive disabilities
- Etiology:
 - Great variability in severity of disability
- Cognitive/mental:
 - Great range and severity of other disabilities



CLINICAL DIAGNOSIS

CEREBRAL PALSY	15
EPILEPSIA	10
SIGN. IMPAIRED VISION	7
AUTISM	4
ADHD	4
GENERAL DEVELOPMENTAL RETARDATION	21
ANSD	2



SYNDROMES

CHARGE	5
DOWN	2
USHER 1	3
WAARDENBURG	5
JERVELL-LANGE-NIELSEN	6
BARTTERS	1
DI GEORGE	1
PENDRED	2
NOONAN	1
JOHANSEN BLIZZARD	1



ETIOLOGY (syndromes excluded)

PREMATURITY	11
ASPHYXIA/PERINATAL	11
CMV	2
CONNEXIN 26	2
MENINGITIS	2
IMMUNODEFICIENCY	1
METABOLIC DISEASE	1

Tests for children with CI

- Little ears
- Ling's sounds
- ESP-N
- Word and sentence recognition tests
- Mullen scales
- Speech evaluation
- TROG



Category scale used in Oslo

10:	≥95% speech perception + including HINT
9:	≥90% speech discrimination
8:	≥70% speech discrimination
7:	≥50% speech discrimination
6:	≥30% speech discrimination
5:	≥ 5% speech discrimination
4:	Vowel discrimination in monosyllables
3:	Vowel discrimination in bisyllables
2:	Perception of phonemes
1:	Perception of speech sounds
0:	Non user

Alternative categories for children with additional disabilities

- 4 Great benefit; adequate speech understanding, verbal communication
- 3 Clear benefit; understands spoken language, uses some words
- 2 Some benefit; reaction to sounds and own voice
- 1 Uses CI, little or no obvious benefit
- 0 Non user

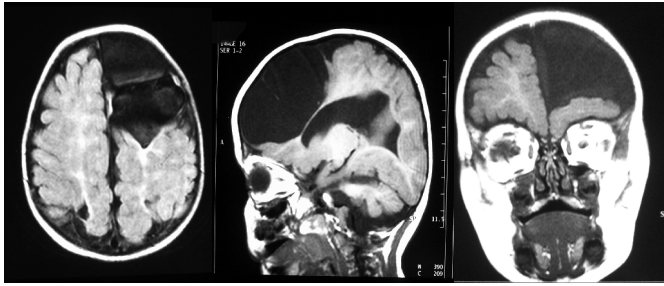
OUTCOME

Category	Score, mean (median)	Range
Alternative category (N=70)	3,0 (3,5)	0 - 4
Speech category (N=27)	7,6 (8)	4 - 9

6 non-users (17-116 months at first CI), 5 mentally retarded

Cognitive disability believed to affect speech development

	YES (N=48)		NO (N=22)	
	Mean (median)	Range	Mean (median)	Range
Age at 1st CI (months)	38 (30)	11 - 118	54 (37)	5 - 150
Follow-up (months)	77 (73)	10 - 165	88 (78)	13 - 204
Alternative category score	2,6 (3)	0 - 4	3,8 (4)	2 - 4
Number in speech category score	10 = 21 %		19 = 86 %	
Nucleus/MEDEL	27 / 21		14 / 8	



Premature (1000 g), CP
13 months, deaf
CI at 2 years 3 months
8 years old: speaks, reads, 94% monosyll. quiet, 56% in noise.



Experiences

- Extensive interdisciplinary management – before and after CI
- Habilitation takes longer time, and more coordination
- Realistic counselling is extremely important
- Screening may reveal deafness *before* other handicaps....



CONCLUSIONS

- CI was beneficial to the vast majority of deaf children with associated disabilities
- Children with cognitive disorders performed worse than those without, but most of them still benefitted from CI
- Generally good results for CP without mental retardation
- Alternative outcome measures covering quality of life needed
- Prognostic factors needed

