YOUNG CHILDREN WITH HEARING LOSS AND ADDITIONAL/ MULTIPLE DISABILITIES

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CHRIB seminar

Introduction

"… I just didn't have the courage to deal with the deafness, because there were so many other things"

- When HL occurs in the presence of co-occurring conditions/ disabilities, the circumstances faced by families and professionals are especially **demanding**

More **challenging** for professionals:
- audiological diagnosis
- amplification
- cochlear implantation
- rehabilitation

Introduction

"… I know that when professionals are working with families of children with disabilities, it can't be easy for them. They have a specialty on which they have to focus, and yet they have a family who is focused on a multitude of things. It is important to remember that families are probably overwhelmed. Their emotions are probably all over the place…"

Mother whose young son has multiple disabilities from CHARGE syndrome
(Roush et al., 2004)

Defining "comorbidities"

A comorbidity is a disease or condition that coexists with a primary disease but also stands on it's own as a specific disease.

Hearing loss often coexists with other disorders, whether presenting as a syndrome or as a constellation of disabilities

Defining "comorbidities"

NDCS, 2012

Defining "comorbidities"

- Often research attributes the high degree of secondary disability in children with HL to **"neurological risk factors"** associated with the cause of the child's HL
- Neurological risk factors are considered to be **medical conditions** that are severe enough to cause a child's deafness, and due to the severity of the condition, often cause additional disabilities for the child (such as vision loss, intellectual disability etc.)

(Szymanski et al., 2012)

Conditions that occur in addition to deafness

- **Gallaudet Research Institute** reported disabilities known to co-occur with hearing loss, based on data from more than 40000 children in the USA (n=42 361)
- Regional and national summary report of data from the *2001-2002 annual survey of deaf and hard of hearing children and Youth.*
  (Washington DC: GRI, Gallaudet University, 2003)
- **40% of children known to have 1 or more disabilities in addition to HL**
- Spectrum of conditions is remarkably diverse:
  "other " conditions = 12.1%

Distribution of conditions that occur in addition to deafness Gallaudet Research Institute, 2003

Condition	% of children n = 42 361
No condition in addition ton deafness	**60.1%**
Learning disability	10.7%
Intellectual disability	**9.8%**
Attention deficit disorder	6.6%
Blindness and low vision	**3.9%**
Cerebral palsy	3.4%
Emotional disturbance	**1.7%**
Other conditions	12.1%

Prevalence of additional disabilities with deafness

- **33-38%** of children with permanent hearing loss have complex developmental needs resulting from secondary disabilities.
  These secondary disabilities = educationally significant
  (Schildroth & Hotto, 1993; Moeller et al., 1990)
- Fortnum & Davids (1997): **38.7%**
- Gallaudet Research Institute (2003) : **40%**

Prevalence of additional disabilities with deafness

NDCS: Prevalence of additional disabilities with deafness: A review of the literature (2012)

- With the advances in hearing screening methods and technology to assess deafness, it is expected previous estimates of prevalence of additional disabilities with hearing loss may no longer be accurate.
- Reviewed published literature after 2002
  (to take into consideration NHS)

Prevalence of additional disabilities with deafness (NDCS, 2012)

Prevalence of additional disabilities in children with hearing loss

- Visual impairment: **4-57%**
- Neurodevelopmental disorder: **2-14%**
- Speech language disorder: **61-88%**

Prevalence of deafness in children with other disabilities

- Autism (ASD): **2-4.2%**
- Cerebral Palsy: **2-13%**
- Pervasive Developmental Disorder: **2%**

Preliminary results of a group of PEDIATRIC CI recipients in South Africa

- **Multi-centric** research study
- **Retrospective** cohort design (retrospective data captured of adult and pediatric CI recipients from patient files & clinical records)
- 6/8 participating CI programs in CI
- **Current sample:** all pediatric (< 18 years) CI recipients of UP CIU & JCIP
- **Sample size** = 287 recipients
  UP CIU: 168
  JCIP: 119

Conditions that occur in addition to deafness in children with CI in SA Bhamjee, Dawood, Noor Mohamed, 2013

Condition	Number of children n = 237	% of children
Visual impairment	**20**	**8.4%**
Mobility impaired	9	3.8%
Cerebral Palsy	**12**	**5.0%**
ADHD	10	4.2%
Autism	**4**	**1.7%**
Emotional/ behavioural disabilities	4	1.7%
Epilepsy	**3**	**1.3%**
Cleft lip and/ or palate	2	0.8%
Apraxia	**4**	**1.7%**
Learning disabilities	5	2.1%

Age of diagnosis of HL: early versus late

Early diagnosis of HL

- Cannot predict outcomes
- Certain disabilities (learning disabilities, mental retardation, ASD) difficult to diagnose in young children – symptoms may not be apparent or can be missed
- Parental expectation
- Counselling NB

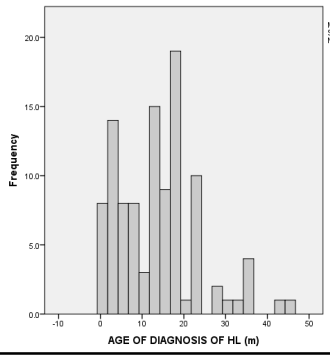
Late diagnosis of HL

- Often, other disabilities diagnosed, but HL not
- Remedial in stead of developmental model of intervention

Age of diagnosis of congenital HL in South Africa

Study	Sample size	Mean age at diagnosis (months)
Venter & Viljoen, 2008	n = 20	31.0
Van der Spuy & Pottas, 2008	n=54	23.0 (SD:18.0)
Swanepoel, Johl & Pienaar, 2013	n= 49	42.1 (SD: 27.6)

AGE OF DIAGNOSIS for congenitally deafened pediatric CI recipients (n=105)

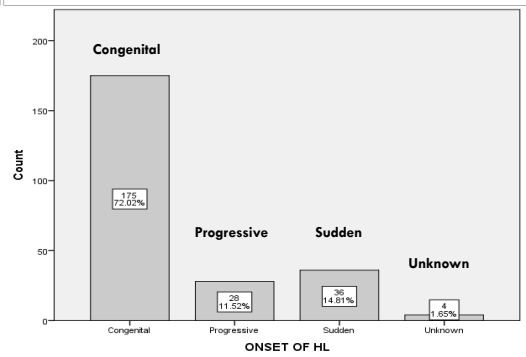


Mean: 13.97 months

(SD = 9.876)

Bjamjee, Dawood, Noor Mohamed, 2013

Preliminary results: Onset of HL (n=243)



DUAL SENSORY IMPAIRMENT IN YOUNG CHILDREN

- When a child has concurrent visual and hearing impairment (some/ complete lack of residual auditory or visual function)
- May be the result of any or a combination of over 70 known causes (such as prematurity, viral infections, Usher and CHARGE syndromes)
- Visual impairment prevalence: 4-59%

⇨ 'visual problems' - 9.5%
⇨ 'significant eye problems' - 12%
⇨ 'need for corrective lenses' - 57%

NDCS, 2012

DUAL SENSORY IMPAIRMENT IN YOUNG CHILDREN

- hearing & vision – **distance senses** (Holte et al., 2006)
  information gathered with these senses are rich & extensive
  (taste, touch, smell = "near senses")
- When either of the distance senses are reduced and yields less information to the child, **learning opportunities** are correspondingly affected. The child's overall development of **cognition** and **communication** are dramatically affected

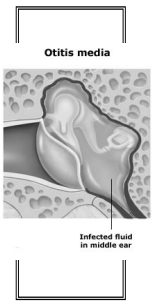
Vision loss limits the degree to which that modality can be used to compensate for the loss of hearing,
whereas the hearing loss limits use of that sensory system to compensate for the loss of vision (Chen, 2004)

DUAL SENSORY IMPAIRMENT IN YOUNG CHILDREN

- Unlike hearing, no universal system for vision screening in newborns
- All children under 3 years of age should have their vision screened, but particularly children with HL
- JCIH Year 2007 Position Statement: *all children diagnosed with hearing loss be referred for a visual acuity screening performed by a paediatric ophthalmologist.*
- Standard pre-school practice: hearing & eye test
- Health professionals should ask parents about or presence of any visual disorders in their young children or their families

(Johnson, 2012)

CO-OCCURING OTITIS MEDIA



- Early childhood conductive HL (as a result of OME) is a pervasive disease that has multiple and potentially longstanding effects on the impressionable young auditory system that may last years after the resolution of the middle-ear fluid.
- It is also associated with significant deficits centring on language skills and verbal tasks as well as behavioural problems, all of which can persists into the teenage years and beyond.

Pillsbury, 2004

CO-OCCURING OTITIS MEDIA

- 1st year of life: more than 50% of children will experience OME, increasing to more than 60% by 2 years of age.
- Although many episodes resolves spontaneously within 3 months, about 30-40% of children have recurrent OME, and 5-10% of those episodes last one year or longer.

The American Academy of Family Physicians, Clinical Practice Guideline: OME, 2004.

Parents and all involved professionals should be advised to be particularly vigilant about OME.

CO-OCCURING OTITIS MEDIA

Middle-ear fluid will likely have **cumulative effects** on a child with SNHL.

The added HL caused by the fluid could mean the difference between **hearing speech and not hearing speech.**

For children with co-existing SNHL's, more **aggressive medical management** for ear disease is warranted to preclude adding to the child's hearing problem and perhaps sabotaging intervention.

Cole & Flexer, 2007

Autism Spectrum Disorder & HL

Szymanski et al., 2012

Prevalence:

- 32 334 deaf and hard of hearing children (2009 – 2010 annual survey of deaf and hard of hearing children and youth – Gallaudet Research Institute)
- 12 595 (39.9%) had an additional disability
- 611 (**1.9%**) diagnosis of ASD & HL
- Gender: ratio approximately 3:1 (male : female)
- 1/59 children (specifically 8 years olds) with HL also receives services for ASD.
- Significantly more children with profound HL had a comorbid diagnosis of ASD than those with milder forms of HL

Autism Spectrum Disorder & HL

Szymanski et al., 2012

Conditions linked to both severe HL and ASD (or characteristics consistent with ASD):

- Rubella
- CMV
- Herpes
- Prematurity
- Toxoplasmosis
- CHARGE
- Meningitis
- Measles

- **Genetics** continues to be implicated in both HL and ASD – both have their own documented genetic influences

HL itself is not a risk factor for behavioural difficulties,

but instead **children who have HL and poor language competence are at the most risk for displaying disruptive behaviour**

Autism Spectrum Disorder & HL

Szymanski et al., 2012

- Documented behavioural interventions, educational strategies and social skills courses for children who are deaf and have ASD do not yet exists.
- Paucity of available literature pertaining to best practice for this population in the areas of intervention, education, diagnosis, language acquisition and general QoL

HL and associated conditions: implications for cochlear implantation

- When deafness is the sole disability: decision on implantation should be based on expected benefits
- What constitutes benefit is still an open question for individuals with additional disabilities.
  - Unclear whether benefit should be considered in terms of speech and language gains or in terms of psychological development and improved quality of life?
  - Presence of additional disabilities – not a contraindication for CI

Berrettini et al., 2008.

HL and associated conditions: implications for cochlear implantation

Minimum objectives for CI in children with multiple disabilities:

1. **Improved auditory awareness** (including the ability to detect voices of family members & other significant individuals)
2. **Improved communication**, supported by access to sound (in the modality most appropriate for the child)
3. **Improved ability to participate in the world around them** through perception of environmental sound relevant for their development and safety

Eisenberg, 2009

IMPLICATIONS: INTERVENTION & EDUCATION

Deaf children with additional needs who may require special adaptation(s) to facilitate learning

- Includes children with specific learning disabilities and ADHD
- Require additional support in the forms of in the form of classroom accommodations, modified teaching strategies, adaptations in academic curricula
- Required accommodations & support compliments those considered appropriate for deaf/ HOH children

Children who are truly multiply disabled

- Combination of 2 or more disabling conditions early in life creates a unique and complex situation
- Methods of intervention and support appropriate for one may not be entirely applicable the presence of the 2nd disability

Eisenberg, 2009

IMPLICATIONS: INTERVENTION & EDUCATION

- Unfortunately, for children with HL and co-occurring conditions, there is a long history of placement decisions based on a categorical view of disabilities

Such an approach fails to consider the uniqueness of each individual

- Can lead to isolated and restrictive placements that focus on the disabilities without fully considering the implications of the child's deafness
- Educational services for deaf students with multiple disabilities are scarce, problematic and not always appropriate

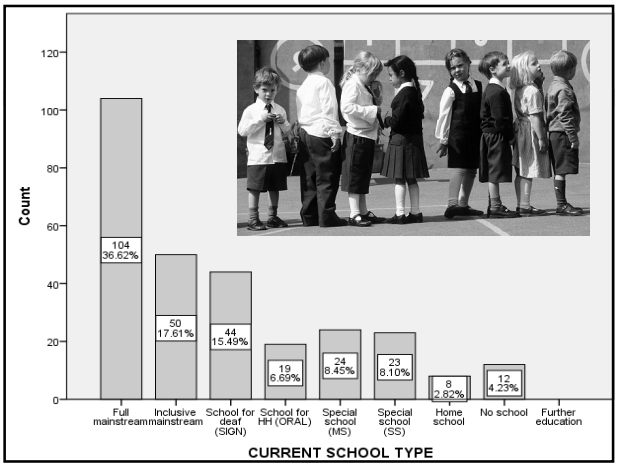
IMPLICATIONS FOR INTERVENTION & EDUCATION

- Meeting the complex educational needs of children with multiple disabilities requires the **involvement of many professionals.**
- The best outcomes are achieved with a **collaborative model** that ensures well-coordinated and integrated planning, placement and intervention.

School placement for a group of pediatric CI recipients in SA (n= 284)

Bjamjee, Dawood, Noor Mohamed, 2013

CURRENTS SCHOOL TYPE	Number of recipients (n-284)	%
Full mainstream	104	36.62%
Inclusive mainstream	50	17.61%
School for the deaf: SIGN medium of communication	44	15.49%
School for hard of hearing: ORAL medium of communication	19	6.69%
Special school: mainstream syllabus	24	8.45%
Special school: special syllabus	23	8.10%
Home school	8	2.82%
No school	12	4.23%



School placement considerations

☆ **School principle:**

- Openness& willingness to learn
- Attitude towards disability (will flow over to staff)

(will be determined by the commitment, assistance and support of involved professionals)

☆ **Small classes**

☆ **Child's role in decision making**

Should spend at least1 day at prospective school

(behaviour will tell a lot)

School placement considerations

4 assumptions to guide the process of school placement:

⇨ Every child is **capable of learning** even when disabilities are multiple and severe

⇨ **Peer acceptance and social relations** are essential for all children, including those with multiple disabilities

⇨ **Families and communities** are critical to success

⇨ Service delivery based on a **trans-disciplinary model** results in coordination of professional disciplines and a sharing of knowledge and resources that results in successful outcomes for families and professionals

Roush et al, 2004

Implications for professionals

⁜ **Multidisciplinary assessment & management**

- a well-coordinated team approach must be established to minimize disabilities, improve outcomes of medical interventions, and initiate appropriate habilitative and educational planning.
- In addition to providing the necessary expertise, a team approach reduces the logistical challenges face by families as they seek developmental assessments from specialists in multiple settings.
- Professionals should be open for each other: learn to trust each other's specialities

Implications for professionals

- Comprehensive **case histories**
- Knowledgeable about the **etiologies** of HL
- Take responsibility for continued **support in school settings**
- **Research:** Growing need for robust published studies assessing the prevalence of additional needs in children with deafness (NDCS, 2012)

IN CONCLUSION

TEAM EFFORT

IN CONCLUSION

For parents:

Take 1 day at a time
Don't worry about tomorrow,
focus on today –
that will change what your choices are tomorrow

"I have learned to trade expectation for appreciation"

Mother of a one year old boy with multiple disabilities

Conclusion

"The best thing a professional can do is to admit that they don't have all the answers, and that no one else does either. Empower families by reminding them that they are the "experts" on their child, because they are with the child everyday and most professionals see them only in the clinic.

Acknowledge their culture, their lifestyles, and be accepting of the ways they cope. And finally, make yourself available to the families when they need you. Remember problems that seem trivial to you are probably huge to families, and with good reason. This is their child, not just another patient. Your patience and understanding is even more valuable than anything you can fix"

Mother whose young son has multiple disabilities from CHARGE syndrome

Roush et al., 2004:343

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