Characteristics of hearing loss and proposal for follow up in children with congenital cytomegalovirus infection

Ina Foulon





cCMV project UZ Brussel



A prospective ongoing study started in 1996

- 1. Obstetrics
- 2. Microbiology
- 3. Neonatology
- 4. Ear Nose and Throat
- 5. Ophtalmology
- 6. Neurology



Obstetrics department:

- Serological screening of all pregnant women followed at the UZ Brussels
 - \rightarrow IgG & IgM at the first prenatal visit
 - → IgG & IgM at birth
- Diagnose time of infection during pregnancy
- Diagnose type of maternal infection
 - → Primary
 - → Non-primary
 - → Unclassified
- Amniocentesis, ultrasound, MRI



Neonatology and Microbiology

- Virological screening of live born infants within 7 days after birth
 - → CMV culture of neonatal urine
 - → CMV culture of neonatal saliva
- Start antiviral medication if necessary
- Diagnose type of infection at birth
 - → Symptomatic: hepatosplenomegaly, petechiae, jaundice, microcephaly
 - → Asymptomatic



Ear Nose and Throat department

- Hearing tests within the first week after birth
 - \rightarrow Clinical examination
 - → Tympanometry (high frequency!)
 - → ABR: Auditory Brainstem Responses
 - → OAE: otoacoustic emmisions
 - → Further hearing test according to follow up



Hearing loss in children with a congenital CMV infection.

Epidemiology

- → Incidence of sensorineural hearing loss in children with congenital cmv-infection, a 10year prospective study. Foulon I, Naessens A, Foulon W, Casteels A, Gordts F. J of Pediatrics. 2008; 153(1):84-8.
- Hearing loss according to the time of maternal infection during pregnancy
 - → Hearing loss in children with a congenital cytomegalovirus infection related to the gestational age at which the maternal primary infection occurred. Foulon I, Naessens A, Foulon W, Casteels A, Gordts F. Pediatrics. 2008 Dec; 122(6):e1123-7.

Characteristics of hearing thresholds

→ Hearing thresholds in children with a congenital CMV infection: a prospective study. Foulon I, Naessens A, Faron G, Foulon W, Jansen AC, Gordts F. International Journal of Pediatric Otorhinolaryngology 76 (2012) 712–717.



Epidemiology: results

- 18.219 mother infant pairs
- 0,61 % of live born children have cCMV
- Type of maternal infection
 - → 50% primary infection
 - → 25% non-primary infection
 - → 25% unclassified
- Type of infection
 - → 91% asymptomatic
 - → 9% symptomatic
- 16% have hearing loss
 - → More frequent in the symptomatic group
 - → More frequent after maternal primary and unclassified group



Time of infection during pregnancy: results

- High incidence of hearing loss in primary infections in the first trimester of pregnancy: 42%
- No hearing loss was observed when infection took place in the second and third trimester of pregnancy



Characteristics of hearing

- Unstable hearing thresholds
 - → Fluctuations
 - → Improvement
 - → Progression/deterioration
 - → Late onset





Universitair Ziekenhuis Brussel



🦾 Universitair Ziekenhuis Brussel





Universitair Ziekenhuis Brussel

Characteristics of hearing thresholds: results

- Need for follow up:
 - → progression took place in 27,3% of the children with SNHL
 - → Late onset hearing loss occurred in 4,3% children with <u>normal hearing at</u> <u>birth</u>
- Improvement of hearing loss in 40,9%
 - \rightarrow Be aware of early intervention
- Important fluctuations (> 30dB) are more related to permanent hearing loss



Overall conclusions

- Children at risk for hearing loss
 - → Symptomatic infection
 - → Maternal primary infection during first trimester of pregnancy
 - → Important (>30dB) changes in hearing threshold



Configuration of hearing loss and retrocochlear involvement and proposal for hearing follow up

Study





Subjects

- Congenitally CMV infected
- Presence of hearing impairment
- Regular audiometric follow up for at least 36 months
- No major other risk factors for hearing loss
- A normal middle ear status





Methods

- Audiological assessment
 - → Revalidation Centre "De Poolster": behavioral testing: CPA/VRA
 - → UZ Brussel (ENT): audiometric evaluation, ABR, OAE, ...
- Audiometry
 - → Most recent audiogram
 - → Most affected ear
 - → 250Hz 500Hz- 1000Hz- 2000Hz- 4000Hz 8000Hz





Results

- 206 children with cCMV
- 22 children with hearing loss (3 14,5 y)
 - → 16%
- 4 exluded
 - \rightarrow 2 are lost for follow up
 - \rightarrow 2 had other risk factors for hearing loss.
- 18 remaining
 - \rightarrow 10 girls , 8 boys



Retrocochlear involvement







Classification of hearing loss



Conductive Hearing loss Sensorineural Hearing loss Retrocochlear hearing loss Cochlear Mixed hearing loss





ABR: Auditory Brainstem responses

Stimulus: click Response measurements, Measuring function of the cochlea and Nv cochlearis and brainstem Measuring thresholds



OAE: Otoacoustic emmisions

- OAE
 - → Measuring function of cochlea
 - → No information about the Nv VIII





Retrocochlear involvement

- An abnormal/absent ABR in combination with present otoacoustic emissions.
 - → Normal cochlear function
- Why is this important?
 - → Necessity of performing ABR in normal hearing children (cCMV)
 - \rightarrow Is the presence of OAE enough to conclude for a normal hearing status?





Retrocochlear involvement in cCMV?

- ABR and OAE within 1 week after birth
- Absent ABR waves and absent OAE
- We never found present OAE
 - → Always cochlear damage

➡ No retrocochlear hearing loss reported.



Conclusion 1

No retrocochlear involvement

- OAE are a useful tool for follow up in normal hearing children
- ➡ Important:
 - ⇒ ABR = labor intensive and often sedation is needed
 - ⇒ Behavioral audiometry: labor intensive and requires a skilled audiologist





Configuration of hearing loss





Cochlea



Audiometry





TOONAUDIOGRAM



Configuration: Presbyacusis







Configuration









Configuration



Ménière



4000 8000 H2 500 1000 2000 - 10 30 12 Hearing Loss (dB) 20 30 40 50 O ÷0 70 80 100 100 R 110 Bone

Otosclerosis

*

Configuration

 Is there a typical hearing configuration in children with cCMV infection?



Results: Configuration

Configuration of most affected ear per subject



Conclusion 2

- - \rightarrow No significant difference for age, gender, ear side
 - → Whole cochlea affected (base and apex)



Other configuration: exclude other etiology



Follow up protocol





Follow up protocol



* Risk for late onset SNHL: 4,3 % ** Risk for progression: 27 %

Note: before hearing evaluation, middle ear disease should be excluded.



cCMV

Thank you.



