

BACKGROUND

- About 1 of every 5 children born with congenital CMV (cCMV) infection will develop permanent problems (such as hearing loss or developmental disabilities) due to the infection.
- Health problems or other disabilities due to cCMV may appear 2 or more years after birth, or they may never appear
 - About 4 of every 5 children with cCMV infection never develop symptoms or disabilities.
- An estimated 15-20 of every 100 cases of bilateral moderate to profound sensorineural hearing loss are attributable to cCMV infection.
- Approximately half of the hearing loss cases associated with cCMV infection can be late-onset. Consequently, they are not detected through a newborn hearing screening program.
- Newborn screening for cCMV infection has the potential to identify infants at risk for hearing loss but currently it is not universally done.
- In 1982, a multidisciplinary study to examine the long-term effects of cCMV was initiated at Texas Children's Hospital and Texas Women's Hospital in Houston.

OBJECTIVE

To describe demographic and audiologic characteristics of infants with cCMV infection enrolled in the Houston Congenital CMV Longitudinal Study.

METHODS

Study population and enrollment

- During 1982-1992 newborns at the Women's Hospital of Texas in Houston were screened for cCMV infection using urine culture obtained during the first 3 days of life and those who were CMV positive were enrolled in the longitudinal study.
- During 1982-2005 newborns with cCMV infection identified elsewhere were referred and included in this study.

Case definitions

- Symptomatic congenital CMV disease:** child with a positive urine culture for CMV within the first 3 weeks of life and one or more of the following signs, symptoms, or laboratory abnormalities: small size at birth, petechiae or purpura (bleeding under the skin), enlarged liver or spleen, jaundice at birth, elevated liver enzymes, low platelet count, small head size, intracranial calcifications, chorioretinitis (vision problems), and hearing loss.
- Asymptomatic congenital CMV infection:** child with a positive urine culture for CMV within the first 3 weeks of life without any above signs.
- Controls:** child with a negative culture for CMV, matched by date of birth to CMV positive infants.

Audiologic testing

- Auditory brainstem response (ABR) test
- Behavioral audiometry test
 - Pure tone average of 500, 1000, 2000 and 4000 Hz were used to calculate degree of hearing loss.
 - Hearing loss degree based on the American Speech-Language & Hearing Association guidelines
- Otoacoustic emissions test (not included here)
- Tests excluded if the tympanograms were abnormal on date of evaluation.

RESULTS

Figure 1. Study population by status

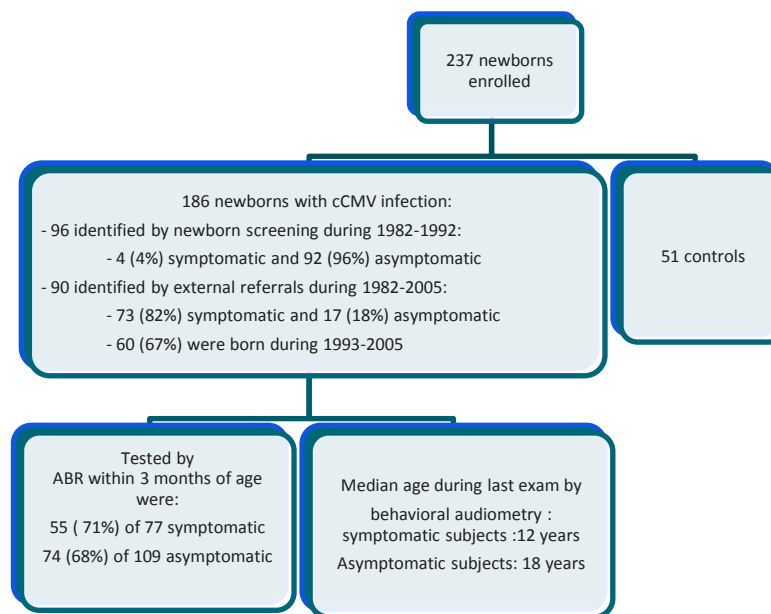


Figure 2. Frequency of hearing loss
Either during first ABR or last exam, children with symptomatic cCMV disease were significantly more likely than children with asymptomatic cCMV infection to have bilateral hearing loss (blue).

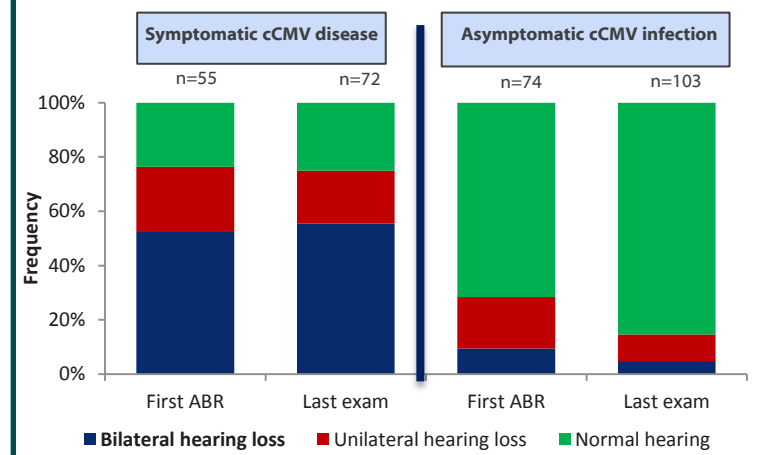
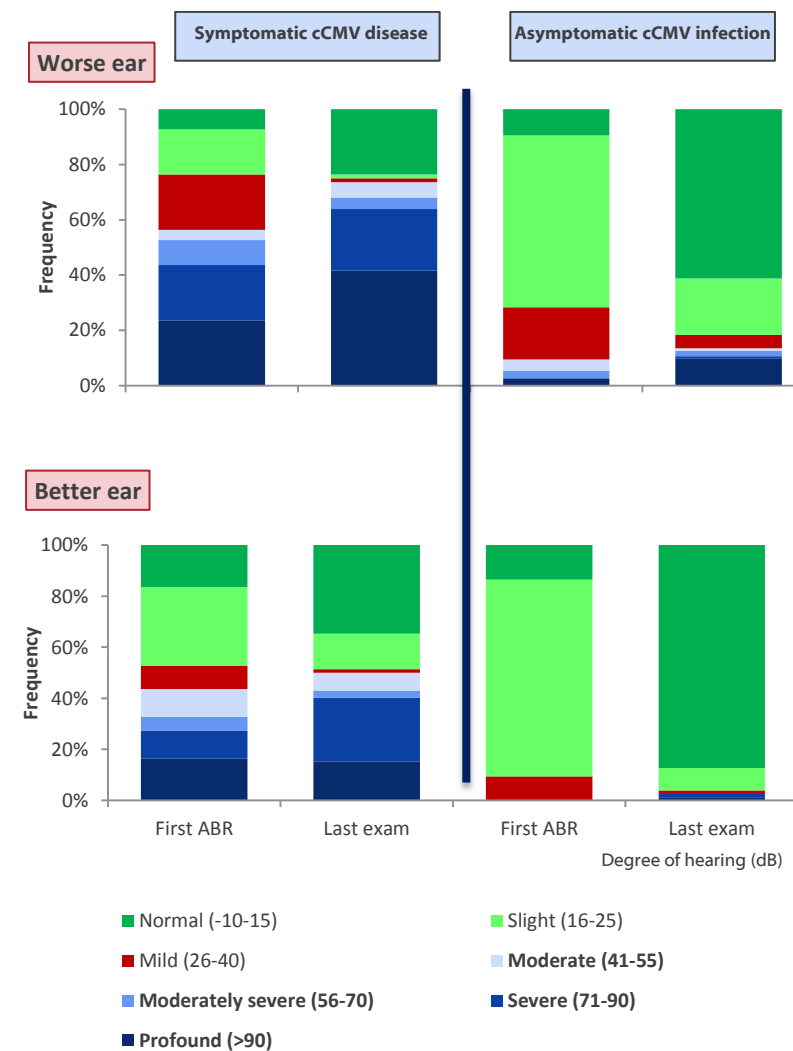


Table 1. Maternal characteristics

Maternal characteristics	Infants with cCMV identified by		Controls n=51 n (%)
	Screening n=96 n (%)	Referrals n=90 n (%)	
20-34 years-old	83 (87)	54 (60)	40 (78)
White race	86 (90)	71 (79)	46 (90)
Non-Hispanic	87 (91)	70 (78)	49 (96)
Married	88 (92)	54 (60)	46 (90)
Private medical insurance	72 (75)	31 (34)	18 (35)
Given birth to more than 1 children	73 (76)	49 (54)	38 (75)

Figure 3. Degree of hearing loss

Children with symptomatic cCMV disease were significantly more likely than children with asymptomatic cCMV infection to have moderate to profound hearing loss (all the blues) at first ABR and at last exam either in the worse or better ear.



SUMMARY

Children with symptomatic cCMV disease:

Within 3 months of age:

- about 50% had bilateral hearing loss, and 25% had unilateral hearing loss;
- moderate to profound bilateral hearing loss was observed in 44% (measured by the degree of hearing loss in the better ear).

At last exam:

- about 65% had bilateral hearing loss (increased from 50%), and 25% had unilateral hearing loss;
- moderate to profound bilateral hearing loss was observed in 50% (increased from 44%).
- After a median follow-up duration of 12 years, a large portion of the symptomatic subjects' hearing loss progressed to a profound degree.

Children with asymptomatic cCMV infection:

Within 3 months:

- Most (70%) had normal hearing.
- 20% had unilateral hearing loss (range mild to profound).
- 10% had bilateral hearing loss (ranging from mild to profound degree in the worse ear, but all mild in the better ear).

At last exam:

- 10% of the unilateral hearing loss subjects' hearing worsened; so only 10% still had unilateral hearing loss (range mild to profound) after over 10 years of follow up.
- 5% of the bilateral hearing loss subjects' hearing worsened; so only 5% still had bilateral hearing loss (mild in the best ear for most children) after over 10 years of follow up.
- Although hearing loss progressed in the asymptomatic group, majority still have relatively normal hearing after median follow-up duration of 18 years

CONCLUSION

- Unilateral, bilateral and progressive hearing losses occurred in both groups, and were a more common occurrence in children with symptomatic cCMV disease than in children with asymptomatic cCMV infection.
- Among children with cCMV identified by newborn screening in our study, approximately 3% had bilateral moderate to profound hearing loss, consistent with other studies reported in the literature.¹
- Although data are not shown here, we have also observed change and fluctuation in hearing among children with cCMV during follow-up, consistent with previous reported studies when children were tracked for 5-6 years.

RECOMMENDATIONS

- Periodic audiologic follow-up of children born with either symptomatic or asymptomatic cCMV is crucial.
- Since hearing loss may occur in otherwise healthy appearing newborns with asymptomatic cCMV infection, newborn screening for congenital CMV infection may detect newborns at risk for hearing loss.

REFERENCE

1- Grosse SD, Ross DS, Dollard SC. Congenital cytomegalovirus (CMV) infection as a cause of permanent bilateral hearing loss: a quantitative assessment. Journal of clinical virology: the official publication of the Pan American Society for Clinical Virology 2008;41:57-62.

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