

CMV, Your Child and Hearing Loss

CMV-induced hearing loss can occur in about half of the children born with symptoms of the virus and in up to 15% of the babies who have a "silent" CMV infection at birth. Regardless of the cause of the hearing loss, any suspected hearing deficit should prompt parents to have a complete hearing evaluation performed expediently. Discovering a loss early may not be easier for the parents, but it will greatly help their child. The news that their child has hearing loss can initially be devastating for the entire family. With the first indication of any loss comes denial, which is frequently followed by guilt, frustration, and possibly anger. It is a normal process that can take some families a few months and even years to work through. However, no matter how much we may fear the truth, it is best to get your child's hearing tested. And of equal importance is follow-up testing, since CMV is notorious for producing a progressive loss (gets worse over time).

Your baby's first hearing evaluation should occur shortly after birth. Sometimes when a diagnosis of congenital CMV infection is made, the child will have a hearing screen done in the hospital nursery prior to discharge. Although it is better to have a hearing screen done on your baby than to have no test at all, a more comprehensive and reliable hearing evaluation called an Acoustic Brainstem Response (ABR) should be performed on your baby before discharge from the hospital or shortly after birth. The types of hearing tests and the

approximate age at which they are to be administered are shown below. Other tests used to assess hearing and other ear functions include tympanograms, acoustic immittance testing, and otoacoustic emissions.

Treatment for sensorineural hearing loss depends on the degree and extent of hearing loss. Depending on the test results, the audiologist may suggest that your child be evaluated by a speech/language therapist.

Speech/language therapists specialize in speech and language development and teach communication skills as well as help your child to maximize their residual hearing. Their goal is to educate, promote independence, improve the quality of life, as well as provide the resources necessary to help your child to reach his or her full potential. They also perform diagnostic evaluations to measure comprehension of speech and if delayed they can provide therapy to help improve your child's language development.

Speech/language therapists also provide information on educational programs and help parents decide what program best meets the child's needs. In addition, they are available to counsel parents on what they can do to help their child benefit most from therapy and to help child to reach their highest potential. It is never too early to begin habitation for a child with hearing loss. As soon as hearing loss is detected, parents need to contact their school district to find out what type educational programs

are available and appropriate. If needed, the speech/language therapist can assist with school placement. Federal law requires schools to provide education for children with hearing impairments. When a hearing impairment has been identified, sometimes the test results indicate that your child could benefit from the use of a hearing device. Listening devices such as hearing aids amplify speech. Your audiologist will discuss the types of hearing devices available and help you decide on the best listening device suitable to meet your child's needs. The types of hearing devices that are available are: hearing aids, tactile aids, FM systems, and cochlear implants. The audiologist can explain the types of devices that are available and help you select the device which will provide the maximum understanding of speech.

To maximize your child's residual senses, it is important to have an ophthalmologic exam performed by an ophthalmologist shortly after birth to rule out any visual problems. According to Dr. Dan Franklin, an otolaryngologist in Houston, Texas, children with hearing impairments need to see well in order to lip read. Optimizing a hearing impaired child's vision will also help them develop good speech and language skills. Hearing loss can cause communication problems and difficulties involving a child's relationship with family, friends, and teachers. The effects of hearing loss can have an impact on your child emotionally and behaviorally.

It is important that you demonstrate acceptance and show support. Be patient. Persons with hearing impairment need a little more time to process speech and sounds. To help build self-esteem, reward your child for his or her accomplishments with positive feedback. It is important that you encourage your child to make friends and interact with others to help prevent isolation. If you and/or your family find that coping with a child who has a hearing impairment is becoming difficult, counseling is available to assist you. Seek advice from your medical professional for a referral to a family counselor. You are not alone in dealing with a child who has a hearing impairment. Support groups and resources are available to help you and your family through the grieving process and help you and your family adjust to living with a child who has hearing loss. The audiologist and speech/language therapist can assist you in finding information and services you need. A good film about a family's experience of dealing with hearing loss entitled, "For a Deaf Son," is available by calling 1-800-368-KERA.

Listed below are names of some other national organizations for hearing impaired persons.

Beginning

P.O. Box 29532

Raleigh, NC 27626-0532

1-800-541-HEAR

Alexander Graham Bell

Association of the Deaf, Inc.

3417 Volta Place, N.W.

Washington, D.C. 20007

202-337-522

| Approximate Age | Hearing Test |
|-----------------|--|
| 0-3 years | ABR (Auditory Brainstem Response) - child must be still, and sedation usually is required - electrodes record brain wave responses to sounds in earphones |
| 3-6 | PA (Pediatric Audiogram) - child wears earphones in a soundproof room - child's responses to speech and sounds are evaluated, often utilizing a game to maintain interest - audiologic assistant is also in the room for help with testing |
| 7+ years | BA (Basic Audiogram) - similar to PA but for older kids - audiologic assistant is not in the room |

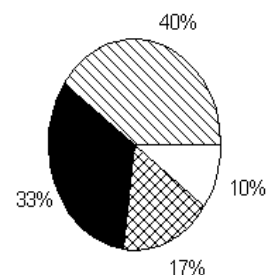
Our special thanks to Susan Williams, an audiologist at Texas Children's Hospital in Houston, TX, for her assistance.

Congenital CMV Infection, a Significant Cause of Hearing Loss

According to current literature, common causes of profound deafness in childhood include genetic disorders (making up over 40%) and meningitis (which contributes up to 10%). That leaves about 50% of childhood deafness due to other causes (such as congenital infections, prematurity, ototoxic drugs, traumatic birth) and unknown causes (Gerber, 1996). One culprit that should no longer be considered an "other" cause of hearing loss is congenital CMV infection. CMV is known to cause hearing impairment in children who

are born with symptoms of the virus. Approximately 38% of these children will have hearing loss at birth and an additional 14% may develop hearing loss during childhood (Demmler, 1997 unpublished data). Considering that 12,000 infants annually are at high risk of developing hearing loss (Northern, 1994), approximately 17% (2,080) can be attributed to CMV. But that's not all. Again, according to Northern, of all infants born with no apparent risk factors for hearing impairment, about 10,800 will develop hearing loss. Congenital CMV infection 90% of the time leaves no apparent signs or symptoms of infection and thus goes undetected in up to 36,000 infants annually. In this group, up to 15% can develop hearing loss, which would result in about 5400 hearing impaired children each year. Therefore, asymptomatic congenital CMV potentially accounts for 50% (5400 or 10,800 deaf children) of hearing loss in children with "no risk factors" annually (Demmler & Littman, unpublished data). From these figures, about 22,800 infants each year will develop hearing loss. Until this time the, "major players" in childhood hearing loss have been genetic causes and meningitis. However, congenital CMV infection should also be in "the majors." About 33% (2,080 + 5400) of all childhood hearing losses annually can be attributed to congenital CMV infection.

Causes of Childhood Hearing Loss



Genetic
 Congenital CMV
 Unknown/Other
 Meningitis

CMV Research Bulletin

What has the CMV Registry been up to ?

When CMV researchers met for the first time in 1990 to try and form the National Congenital CMV Disease Registry, there were 35 interested physicians. Since that time the Registry has grown over 45% and now has over 50 participating physicians and researchers.

Additionally now we have participants in 24 states, which is up from 16 states. The two newest members and locations are Dr. Fiedler in Camden, New Jersey and Dr. Nelson in Lexington, Kentucky. **Welcome !!**

Since its inception, the CMV Registry has been not only a growing body of information on infants born with symptoms of CMV infections, but it has also been a tool for collaborative research. We welcome every opportunity to learn more about this virus we so simply call CMV. Therefore, one of the goals of the CMV Registry has been to promote collaborative efforts among researchers. Over the years we have shared our findings, insights, and data with researchers who have used the information as a reference and/or as a subset of their own study population. Investigators and topics the Registry has been involved with include "Exploring the role of HLA-G as an antigen presenting molecule to T-cells of mother who give birth to CMV-infected infants" by Dr. Riddell in Washington; "Congenital CMV disease in the Louisville area" by Dr. Marshall in Kentucky; "Seizures and congenital CMV infection" by Dr. Bale in Iowa; and "Congenital CMV infections in twins" by Drs. Rollin and Stewart at the Centers for Disease Control and Prevention (CDC) in Atlanta. Each of these investigators have worked with CMV Registry data

and have published or presented their findings. We wish to thank all of these physicians for including data from the CMV Registry in their research and we also would like to encourage others to consider collaborative efforts.

Facts & Figures

As of April, 1997, the CMV Registry has recorded 581 cases of symptomatic congenital CMV infection. At this time we are a bit behind the previous years as far as the number of cases received goes. We know this is not likely due to a decrease in the number of affected babies, so participants, get those forms filled out and send them to us.

We are preparing another summary of our findings for the year 2000! To have the most complete information on each case report, we are still planning to send each participant a list of cases submitted thus far. The most frequently overlooked items on the questionnaire are:

| Characteristic | % Unknown |
|--|-----------|
| Mother's age | 3 |
| Marital status | 8 |
| Mother's ethnicity (Hispanic?) | 25 |
| Platelet count equal to/less than 75,000/mm ³ | 8 |
| Neurologic abnormality | 11 |
| Direct bili equal to/greater than 3 mg/dl | 15 |
| Intracranial calcifications | 15 |
| Chorioretinitis | 22 |

| | |
|--------------------|----|
| ALT > 100 IU/L | 27 |
| Hearing impairment | 38 |

Top Ten Reporting Centers

The top ten reporting centers are listed in the following table.

| | |
|----------------------|----------|
| Dr. Pass/Dr. Bopanna | 80 |
| Dr. Demmler | 60 |
| Dr. Dankner | 46 |
| Dr. Kumar | 37 |
| Dr. Brady | 29 |
| Dr. Reuman | 27 |
| Dr. Ford-Jones | 26 |
| Dr. Gruber | 25 |
| Dr. Shelton | 24 cases |
| Dr. Chonmaitree | 20 cases |

As you can see, the #1 reporting center is the University of Alabama at Birmingham (UAB) with Drs. Pass and Bopanna. Since the Registry began, this group has consistently reported more cases of symptomatic congenital CMV infection than any other physician or group of physicians. Does Alabama have more CMV than other parts of the country? The answer to this question is most likely *No*. Alabama, or more specifically, Birmingham, does not appear to have a greater prevalence of congenital CMV infection compared to other cities of similar size and demographics. Similar to the rest of the country, about 1% of all births in Birmingham have CMV and about 6% of these have symptoms at birth. What are different at UAB, however, are the methods used to

capture their figures on the infection rate. For many years Dr. Pass and colleagues have been screening live births at several hospitals within the city for congenital CMV infection. By testing a newborn's urine or saliva, they can detect congenital infection regardless of the baby's clinical signs or symptoms (whether or not the baby has any symptoms of the virus). Therefore, UAB detects more babies with symptomatic congenital CMV infection because they are "actively" searching for them and they are also able to produce an accurate infection rate for their area.

To be added to our mailing list, please send your request to us by the Subscription Form, or by postal mail or e-mail (addresses follow).

Please send correspondence to:

*National Congenital CMV
Disease Registry
Feigin Center, Suite 1150
1102 Bates Street, MC 3-2371
Houston, TX 77030-2399
Phone: 832-824-4387
FAX: 832-825-4347
cmv@bcm.edu*

More Info...

E-mail Bag

Members of the CMV parent-to-parent support network, it is time to

update the information on your listing. We will send you a new consent form in the near future. Please take a moment to complete the form and send it back to us. We are also asking that if you have Internet access please share with us your electronic mail (e-mail) address to facilitate communication between parents.

An e-mail CMV listserv is now available!! The list was created and is maintained by Sue Robertson, mother of Ronda (Roo). She can be contacted at her E-mail address (peacock351@aol.com). By subscribing to the list parents can share their trials and triumphs, while others may join to simply to learn more about congenital CMV infection through this discussion format. Everyone interested is welcome. To subscribe, send the following command to Majordomo@avenza.com. In the **BODY** of the e-mail message type: subscribe CMV-L.

Support for parents

As many of you already know, there is a CMV parent-to-parent support network. The members and list are currently maintained at the CMV Registry. If you are a parent of a child who has CMV or if you are a therapist or educator who would like to help a family affected by CMV, please call, write, FAX, or e-mail us. We will send you the consent form to fill out. Currently we have 92 families who have joined from the USA and one family

from Japan. We also hope to have some families from Canada on our list soon.

CMV Quiz

True or False Children with CMV should not swim or receive therapy in public pools where other children and pregnant women may be swimming?

False. This virus is easily inactivated by contact with the outside surroundings, therefore in a swimming pool that is well maintained at the correct chlorine and pH levels, CMV should be inactivated. Furthermore, the dilutional effect of a small amount of saliva or urine entering a pool would make transmission of even a live virus highly unlikely.

True or False A baby who catches CMV during delivery or from breast milk is at risk for problems (developmental delays, hearing loss, etc.) just like babies with congenital CMV infection?

False. The long-term effects of congenital CMV infections are most often related to injury of the central nervous system (CNS) that occurred while the baby was developing. Therefore infants infected during or after delivery are not affected in the same way and they usually have a symptom-free infection. However, premature infants and infants with underlying conditions who catch CMV very early in life, may experience express significant symptoms of the infection.

NEWSLETTER SUBSCRIPTION AND PARENT SUPPORT NETWORK INFORMATION
MAILING ADDRESS CHANGES
CMV RESEARCH DONATIONS

- I would like to be (added to / deleted from) the CMV Updates mailing list.
- I would like to be added to the CMV Updates email list.
- I have an address change.
- I would like to receive information about the congenital CMV disease Parent-to-Parent Support Network.
- I would like to be deleted from the Parent-to-Parent Support Network.
- Enclosed is my \$_____ donation to continue research on congenital CMV disease and infection. Please make checks payable to the "CMV Research Fund," which is affiliated with Baylor College of Medicine and Texas Children's Hospital, Houston, Texas. All donations are tax deductible.

Name: _____

Address: _____

Phone: (_____) _____

What is your interest in CMV infection?

- Parent/Family member of a child with congenital CMV disease or infection.
- Health care professional. Specify: _____
- Other. Specify: _____

Detach and mail this form to:

CMV Registry, Feigin Center, Suite 1150 • 1102 Bates Street, MC 3-2371 • Houston, Texas 77030
 Telephone: 832-824-4387 • Fax: 832-825-4347 • E-mail: cmv@bcm.edu